

**The psychosocial adjustment of children
with major congenital abdominal anomalies**

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**The psychosocial adjustment of children with
major congenital abdominal anomalies**

De psychosociale aanpassing van kinderen met ernstige aangeboren darmafwijkingen

Proefschrift

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Abbreviations

ADQC	Abbreviated Depression Questionnaire for Children
ARM	Anorectal malformations
AWD	Abdominal wall defects
CAA	Congenital abdominal anomalies
CBCL	Child Behavior Checklist
CD	Crohn's disease
CDH	Congenital diaphragmatic hernia
CFA	Cranio-facial anomalies
CHD	Congenital heart disease
CHQ	Child Health Questionnaire
CQOL	Child Quality of Life
EA	Esophageal atresia
ECMO	Extracorporeal membrane oxygenation
FAD	Family Assessment Device
FDI	Functional Disability Inventory
HD	Hirschsprung's disease
HRQOL	Health-related quality of life
IA	Intestinal atresia
IBD	Inflammatory bowel disease
IPSQ	Interpersonal Problem Solving Questionnaire
IQ	Intelligence quotient
KIND-L	Munich Quality of Life Questionnaire
LEQ	Life-Event Questionnaire
PPS	Play Performance Scale
QL	Quality of life
QLQC	Quality of Life questionnaire for Children
QWB	Quality of Well-Being scale
RAP	Recurrent abdominal pain
SES	Socio-economic status
TACQOL	TNO-AZL Child Quality of Life
TRF	Teacher's Report Form
UC	Ulcerative colitis
VABS	Vineland Adaptive Behavior Scales
WISC-RN	Wechsler Intelligence Scale for Children - Revised Nederlandse vertaling



Chapter 1

Background of the study

1.1. Introduction

The confrontation with the birth of a child with a congenital anomaly is enormously stressful for parents. Expectations of a healthy baby and a joyful nursery time are often not met. These parents will have multiple questions and concerns which can be centered around the past, the present, and the future. Looking back, they may ask themselves: What has gone wrong that our child has a congenital anomaly? What have we done or not done that this has happened? In most cases these questions cannot be answered, because no specific cause of the anomaly can be identified. This often leaves parents with much uncertainty, which will not easily be resolved. At the same time, they will have to accept the situation as it is, and face the responsibility to care for this child.

This presents the parents with the actual task of caring for and attaching to their newborn child. However, these children must be hospitalized, often in a neonatal pediatric surgery intensive care unit during a considerable time, which aggravates the difficulties for the parents. The unit in itself may provoke anxiety in the parents. They find themselves in a completely unfamiliar situation of which they do not know the rules and what is expected of them. They see their child surrounded by high-tech instruments on a ward with many busy doctors and nurses. The child will be undergoing many more or less invasive and painful procedures, and it may be very difficult for parents to see this happen, while they have very few tools to comfort or help the child. The medical situation of a child with a major congenital abdominal anomaly will be critical for a shorter or longer period. For parents this is a situation which provokes enormous anxieties. As a consequence they may have difficulty to attach themselves to their child until the medical situation has been stabilized.

If a child with a major congenital abdominal anomaly has survived these first months, concerns about the future may arise. Parents will raise questions about what will become of their child. Will it grow up to be a normal child? Will it be handicapped? In what ways will the life of their child be affected by the anomaly or the treatment? These questions are often very difficult to answer for a particular child, and answers based on group findings may be unsatisfying for parents and may not take away their uncertainties.

So, during the first year after the birth of a child with a major congenital abdominal

anomaly many parents are confronted with numerous questions, stresses, and uncertainties. Medicine not only has a task to treat these children in an adequate way but also to support the parents and help them cope with these problems. One way to do this, is to provide parents with answers to their questions based on well-documented information.

For the team of pediatric surgeons and nurses who treat children with a major congenital abdominal anomaly, the situation is by no means always unambiguous. Surely, they are confronted with children whose anomalies may be severe, but who have a good prognosis even if treatment may be difficult and prolonged. But on the other hand they are confronted with children who may have multiple anomalies, who may be at risk for neurocognitive damage, and who may need very lengthy intensive care treatment. For some of these children, questions may be raised about the appropriateness of treatment. Are we treating a child whose future functioning may be severely limited? Will this child in the future be confronted with major handicaps and will it suffer emotional damage due to the long hospitalization and its consequences? In these situations prolonging treatment might mean only prolonging suffering without the perspective of future acceptable quality of life. This confronts physicians and nurses with serious ethical dilemma's, which cannot be properly discussed without a good insight in the prognosis of these children and in the factors influencing the prognosis.

Pediatric surgeons are not only confronted with the medical problems of children with congenital abdominal anomalies in their newborn period, but they follow these children often into adulthood. During these follow-up visits they are confronted with a large diversity of outcomes. Some children are doing rather well and have only minor problems. Other children, however, have more serious physical problems. Pediatric surgeons identified especially children with Hirschsprung's disease and anorectal malformations as being at risk for psychosocial adjustment problems, because they often have problems with defecation and fecal continence which may lead to shame and embarrassment. Because pediatric surgeons lack the tools to evaluate the nature and extent of these emotional and social problems during hospital visits, they could not verify these concerns. Because, as indicated before, the burden for parents and families with a child with a congenital anomaly may be very heavy, concerns are also raised how these anomalies affect future family and parental functioning. So, more insight into the emotional and social functioning of children with congenital abdominal anomalies and of their families is needed in order to come to better recognition of children at risk for maladjustment, of the factors which may have a major influence on outcome, and the possibilities to provide guidance for these children and their families.

Many of the aforementioned questions might be answered based on previous research, while other questions need additional empirical research. To explore the available empirical

evidence, the literature concerning the psychosocial functioning of children with congenital anomalies in general and congenital abdominal anomalies in particular and its determinants will be briefly reviewed.

1.2. Psychosocial functioning of children with congenital anomalies

The most important anomalies under study are congenital heart disease (CHD), craniofacial anomalies (CFA), neurological disorders (myelomeningocele, spina bifida), urogenital, and abdominal anomalies.

Children with CHD were found to have lower intelligence scores than children in the general population and more feelings of anxiety and inferiority (Kramer, Awiszus, Sterzel, Van Halteren, & Claßen, 1989). Increased emotional problems for children with CHD were reported by parents (Casey, Sykes, Craig, Power, & Mulholland, 1996; Utens et al., 1993). In a recent study on overall quality of life in children with CHD, however, no differences between these children and children from a comparison group were found (Moyen-Laane et al., 1997).

In children with craniofacial anomalies (CFA) whose physical appearance is sometimes seriously affected, much research has been directed at the assessment of self-esteem and social skills. Children with CFA experienced increased social problems such as being teased, social withdrawal, and shame (Kapp-Simon & McGuire, 1997; Kapp-Simon, Simon, & Kristovich, 1992; Pruzinsky, 1992). Pope and Ward (1997) found that self-perceived facial appearance was associated with problems in peer relations and low global self-esteem. Social skills, however, appeared to be in the normal range (Kapp-Simon et al., 1992). Overall psychosocial adjustment as reported by parents was found to be decreased for children and adolescents with CFA compared to normative data (Kapp-Simon et al., 1992; Padwa, Evans, & Pillemer, 1991).

Children with congenital neurological anomalies showed a wide range of physical, psychological, and social problems at follow-up. Moderate to severe deficits in motor function, visual and auditory function, and urinary continence were reported for children with hydrocephalus (eg. Hoppe-Hirsch et al., 1998) or children with spina bifida (eg. Wallander, Feldman, & Varni, 1989). Cognitive impairments were described for children with spina bifida and/or hydrocephalus (Hoppe-Hirsch et al., 1998; Wills, Holmbeck, Dillon, & McLone, 1990). Finally, children with spina bifida and especially those with hydrocephalus showed higher rates of psychosocial maladjustment (Fletcher et al., 1995; Wallander et al., 1989), and lower self-esteem (Appleton et al., 1994; Fletcher et al., 1995).

Very few studies are devoted to children with urogenital anomalies. Mureau, Slijper, Slob, and Verhulst (1997) did not find increased maladjustment for children and adolescents with hypospadias. Other studies, however, did find more behavioral and emotional problems in

hypospadias patients (Blotchky & Grossman, 1979; Lepore & Kesler, 1979; Sandberg, Meyer-Bahlburg, Aranoff, Sconzo, & Hensle, 1989). Diseth, Bjørdal, Schultz, Stange, and Emblem (1998) did find high rates of psychiatric disturbance in adolescents with epispadias and bladder extrophy (50 % in study population versus 12 % in controls). Surprisingly however, they did not find increased maladjustment using standardized questionnaires concerning emotional and behavioral problems. In conclusion, for urogenital malformations the evidence of increased psychosocial problems at follow-up is at least conflicting.

There are a few studies indicating that children with congenital abdominal anomalies (CAA) show cognitive, emotional, and behavioral problems. Children with congenital diaphragmatic hernia are at risk for developmental delays (Davenport, Rivlin, D'Souza, & Bianchi, 1992; Nobuhara, Lund, Mitchell, Kharasch, & Wilson, 1996). Cognitive problems were reported for children with esophageal atresia (Dera, Mies, and Martinus, 1980). Increased emotional problems for children with esophageal atresia were reported by Dera et al. (1980) but overall quality of life in adult populations appears to be in the normal range (Chetcuti, Myers, Phelan, and Beasley, 1988; Lehner, 1989; Ure et al., 1998). Increased emotional and behavioral problems were reported for children with abdominal wall defects and anorectal malformations (Diseth & Emblem, 1996; Ginn-Pease et al., 1991; Ludman, Spitz, & Kiely, 1994).

It can be concluded that across different congenital anomalies there is an increased risk for psychosocial problems, such as emotional and behavioral problems and lowered self-esteem, while the presence of cognitive and learning problems are reported for some congenital anomalies, especially neurological and cardiac anomalies.

1.3. Determinants of psychosocial adjustment of children with congenital anomalies

A major question in the follow-up of physical disorders is which are the determinants of psychosocial adjustment to physical disease. There is a broad body of research which has led to some important conclusions. There appears to be very little effect of type or severity of acquired or congenital physical disorders on psychosocial functioning of children (Wallander & Varni, 1998). The most prominent difference in effect-size on overall adjustment over a large number of studies was found for sensory and neurological disorders as appeared from a meta-analysis by Lavigne and Faier-Routman (1992). More important influences on the level of psychosocial adjustment for congenital or acquired childhood physical disorders appear to be factors related to the parent-child relationship, family functioning, and psychosocial functioning of the parents. The psychosocial functioning of children with congenital anomalies has been related to aspects of family functioning such as level of conflict and control (Murch & Cohen, 1989), family cohesiveness (Lavigne, Nolan, & McLone, 1988; Varni, Rubenfeld, Talbot, & Setoguchi, 1989),

and parenting stress (Krueckeberg & Kapp-Simon, 1993). Only one study addressed the psychosocial functioning of children with a congenital abdominal anomaly in relation to family functioning (Diseth, Bjørland, Nøvik, & Emblem, 1997). In this study parental warmth was the strongest predictor of global psychosocial functioning of children with Hirschsprung's disease.

1.3.1. Parental and family functioning of children with congenital anomalies

Studies on parental and family functioning of children with congenital anomalies pertain to attachment, family functioning including marital relationship, and parental psychosocial functioning.

1.3.1.1. Attachment

As stated above, there are ample reasons to expect a more problematic attachment between parents and children with congenital anomalies. However, based on a meta-analysis of 34 clinical studies, Van IJzendoorn, Goldberg, Kroonenberg, and Frenkel (1992) concluded that maternal problems such as mental illness are more related to deviant attachment than child characteristics such as prematurity or deafness. Several studies concerning the attachment between children with CFA and CHD and their mothers support these conclusions (Goldberg, Washington, Morris, Fischer-Fay, & Simmons, 1990; Hoeksma, Koomen, & Koops, 1987; Speltz, Endriga, Fisher, & Mason, 1997). No studies are available on attachment in congenital neurological, urogenital, or abdominal anomalies.

1.3.1.2. Family functioning and marital relationship of the parents

Very few empirical studies concerning family functioning are available for children with chronic conditions as well as with congenital anomalies. Wallander and Varni (1998) stated that it is difficult to come to a coherent conclusion based on the sparse research concerning functioning of families with children with chronic disorders. There are some studies that have assessed family functioning in families of children with congenital anomalies. Heller, Rafnan, Zvagulis, and Pless (1985) found an equal proportion of children with CHD, CFA, and hearing defects coming from low (i.e. problematic) functioning families as children in the normal population. Fletcher et al. (1995) did not find worse family functioning for children with hydrocephalus compared to children from the normal population. Diseth reported no increased family difficulties for children with urogenital anomalies (Diseth et al., 1998) or Hirschsprung's disease (Diseth et al., 1997).

In a review of the literature on the effect of having a congenitally handicapped child on the marital dyad Benson and Gross (1989) concluded that divorce rate, as a gross measure of the quality of the marital relationship, is not higher in these populations than in the general population. Further, they concluded that there may be positive as well as negative influences of

having a congenitally handicapped child on marital satisfaction (Benson & Gross, 1989). Some recent studies indicated that the marital quality of parents of children with spina bifida is not worse compared to the general population. Kazak, Segal-Andrews, and Johnson (1995) concluded that there is strong evidence that there are no differences in overall rates of marital satisfaction between families with and without physically affected children.

The conclusion seems justified that family functioning as a whole and quality of the marital relationship in particular are mostly normal in pediatric populations with congenital anomalies.

1.3.1.3. Parental psychosocial functioning

There is a wealth of studies on the maternal adjustment to having a child with a chronic disease. These studies showed that mothers of chronically sick children do have more mental health problems, but that figures may be exaggerated if these are solely based on clinical populations compared to general population based studies (Eiser, 1993). Several recent studies on samples of children with congenital anomalies indicated that parental psychosocial functioning is not necessarily worse in these situations. Holmbeck et al. (1997) and Capelli, McGrath, Daniels, Manion, and Schillinger (1994) found that mothers of children with spina bifida did not report more psychological problems, although Holmbeck et al. (1997) did report more psychological problems for the fathers. Campis, DeMaso, and Twente (1995) reported no increased depression or anxiety for 77 mothers of children with CFA. Based on several studies Wallander and Varni (1998) concluded that type of illness or disability, nor severity of disability are associated with maternal adjustment.

From this review it can be concluded that children with congenital anomalies are at risk for later psychosocial problems, and that environmental factors such as family functioning appear to play an important role in the outcome of the children. However, family and parental functioning in itself are not necessarily more problematic in families with a child having a congenital anomaly than in the general population. Concerning the follow-up of children with congenital abdominal anomalies there is still a great lack of sound research. Critique on the existing literature can be summarized as:

1. Not all major congenital abdominal anomalies have been studied. Especially, very few data are available concerning diaphragmatic hernia, intestinal atresia, and abdominal wall defects.
2. Most research in this area concerns future physical functioning while only a minority of studies concern psychosocial functioning.
3. Many of the follow-up studies on congenital abdominal anomalies concern for a large part adult populations, while little is known on pediatric populations.

4. In most studies non-standardized assessment procedures have been used. This makes comparisons of psychosocial functioning of this group with the general population or normative data difficult.

Because very few answers to questions of parents and doctors of children with major congenital abdominal anomalies can be answered based on existing literature pertinent to these anomalies, it was needed to plan a follow-up study on the physical, but especially, the psychosocial functioning of these children.

1.4. Aims of the study

The design of the present study was guided by the questions raised above. These questions can be summarized as:

1. How is the physical, psychological, and social functioning of children who are born with a major congenital abdominal anomaly when they reach middle childhood?
2. Which medical parameters have an important influence on the outcome, especially concerning psychosocial functioning, of these children?
3. Which other, especially environmental, factors are related to the future psychosocial functioning of these children?
4. Which children can be identified who run the greatest risk for problems in their psychosocial functioning and as such have the greatest need for psychosocial guidance or support?

The theoretical perspective of a part of this study was based on the quality of life paradigm. In the realm of physical disorders this is usually referred to as health-related quality of life (HRQOL). Spieth and Harris (1996) discern two main conceptual models of HRQOL, the utility model developed among others by Kaplan and Anderson (1988), and the health status measurement model. The health status measurement approach has been adopted in the majority of HRQOL research in pediatric populations (Spieth and Harris, 1996). Starting from the definition of the World Health Organization of health as a state of complete physical, mental, and social well-being, there is general agreement that HRQOL is a multidimensional construct including three broad domains, i.e. the physical, psychological, and social functioning domains (Aaronson, 1991; Kaplan, 1988; Spieth & Harris, 1996). The necessity to assess the HRQOL of children in those three main domains is acknowledged by most researchers, although there is considerable variability in the choice of subdomains. The physical functioning domain generally includes disease state or symptomatology and functional status, i.e. the ability to perform age-appropriate daily activities (Spieth & Harris, 1996). There is no clarity in the literature on

HRQOL measurement concerning the subdomains which should be included in the psychological and social functioning domains. Although the need for self-report HRQOL instrument is stressed by several authors (Eiser, Havermans, Craft, & Kernaham, 1995; Mulhern et al., 1989; Rosenbaum & Saigal, 1996), to date, only two self-report HRQOL instruments have been published, the Munich Quality of Life Questionnaire (Bullinger, Von Mackensen, & Kirchberger, 1994) and the Child Quality of Life Questionnaire (Graham, Stevenson, & Flynn, 1997).

1.5. Design of the study

In this study a large non-selected group of children with different major congenital abdominal anomalies were investigated. Based on the HRQOL paradigm, a broad range of standardized assessment instruments concerning the physical, psychological, and social functioning of the children were used. To be able to assess the health-related quality of life of these children specifically, a generic self-report and parent-report HRQOL instrument was developed. To identify determinants of the HRQOL of these children not only background physical parameters but also parental and family functioning were assessed using reliable measures.

1.6. Structure of the report

In chapter 2, the literature concerning the quality of life of children with abdominal disorders in general and congenital abdominal disorders in particular will be reviewed. In chapter 3, an overview will be given of the study samples and the assessment instruments. In chapter 4, the development and psychometric properties of a HRQOL instrument for children will be described. In the next three chapters the psychosocial functioning of children from specific diagnostic categories will be described: congenital diaphragmatic hernia in chapter 5, esophageal atresia in chapter 6, and Hirschsprung's disease and congenital anorectal malformations in chapter 7. In chapter 8, the results concerning the psychosocial adjustment of the whole group will be reported. In the latter chapter, the psychosocial adjustment of the children will be brought into relation with medical and psychosocial background variables from the perspective of the so-called disability-stress-coping model of Wallander and Varni (1992). In chapter 9, the study will be summarized, conclusions will be drawn, and recommendations concerning clinical practice and future research will be given

Chapter 2

Quality of life in abdominal disorders

Nico Bouman

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2.1. Introduction

Abdominal disorders consist of rather diverse disorders with equally diverse consequences for quality of life (QL). On the one hand there is recurrent abdominal pain (RAP), a common problem affecting some 10 % of all children. Although RAP is a disorder with little organic pathology, it raises much concern with parents and it may influence the QL of a large proportion of children. On the other hand there are the inflammatory bowel diseases (IBD), which are rare diseases with severe morbidity affecting QL in several domains. The congenital abdominal anomalies fall somewhat outside the scope of the former conditions. They are not diseases in the strict sense, but may nonetheless have long-term consequences on the health and QL of children who were born with these anomalies.

Research on the QL of abdominal disorders in childhood and adolescence in strict sense is scarce, although several studies have been performed focusing on the physical and psychological functioning of RAP, IBD, and congenital abdominal anomalies. Very few QL instruments have been developed for these age groups. In this chapter, QL is defined as a multidimensional construct including the physical, psychological, and social functioning domains (Aarons, 1991; Spieth & Harris, 1996). The physical functioning domain generally includes disease state or symptomatology and functional status, i.e. the ability to perform age-appropriate daily activities (Spieth & Harris, 1996). There is no clarity about the issue which aspects of psychological and social functioning should be included in QL measurement. Inclusion of overt behavioral problems in the psychological functioning domain is recommended by Spieth and Harris (1996). Other aspects of psychological functioning such as cognitive functioning and self-esteem will be described. In the social functioning domain relations with peers, with family

members, and teachers are primarily identified as the most important aspects and will be discussed (Spieth and Harris, 1996).

This review will focus on what is known about the physical, as well as the psychological and social consequences in childhood and adolescence of these disorders. In the first part, RAP will be described, in the second part, IBD, and in the last part, congenital abdominal anomalies. Each part will describe the medical aspects of the disorder, followed by the description of the physical functioning and the psychological and social functioning, and will end with a brief discussion of possible future QL research.

2.2. Recurrent Abdominal Pain

RAP is a common problem during childhood. It is defined as at least three episodes of abdominal pain affecting the activities of the child over a period of more than three months (Apley, 1959, 1975). The reported prevalence of RAP among school-aged children varies from 5 % to 25 % (Faulk & Nicol, 1986; Stevenson, Simpson, & Bailey, 1988). Hyams, Burke, Davis, Rzepski, and Andrulonis (1996) found that 21 % of a community based sample of 507 adolescents had weekly complaints of abdominal pain severe enough to interfere with activities. Longitudinal research indicates that RAP is a chronic condition for a part of the children. In a follow-up study of their original groups of patients, Apley and Hale (1973) showed that abdominal pain is persistent into adulthood in one third of cases. In a follow-up study of a cohort of 136 4-year-old children until the age of 10, Borge, Nordhagen, Moe, Botten, and Bakketeig (1994) found a strong relationship between the number of complaints at the age of 4 and at the age of 10. Fifty-five percent of children with frequent stomachaches at the age of 4 still had these complaints at the age of 10, while only 13 % of children without stomachache at the age of 4 had frequent stomachaches at the age of 10. Finally, Walker, Garber, Van Slyke and Greene (1995) reported persistent high levels of abdominal pain in a group of 31 children with RAP 5 to 6 years after diagnosis.

Only in about 10 % of cases with RAP an organic cause is found (Apley, 1959). Based on a study of 106 children with chronic stomachache in which 42 % of children had organic causes for their complaints, Van Der Meer (1993) argues that this is an underestimation and that more efforts should be made to find an organic cause in a child presenting with RAP. However, these results may be explained by the fact that this study concerned only clinically referred children, a part of which were hospitalized, which may represent a more serious subgroup of children with RAP with a greater chance to find organic causes.

Treatment of RAP consists of avoiding invasive medical procedures in the first place. Apley and Hale (1973) recommend explanation, reassurance, and discussion with the child and its parents by the treating physician. Finney, Lemanek, Cataldo, Katz, and Fuqua (1989) and Sanders, Shepherd, Cleghorn, and Woolford (1994) describe brief structured therapies primarily based on cognitive-behavioral techniques as successful treatment strategies.

2.2.1. Physical functioning

By definition children with RAP suffer from frequent abdominal pains. Other common complaints are nausea, vomiting, and food intolerance (Hyams et al., 1996; Van Der Meer, 1993). Although, as included in the definition, pain should interfere with daily activities of the child, it is not clear in most studies to what extent children with RAP are limited in their activities. In the study by Hyams et al. (1996), 51 % of adolescents with abdominal pain missed at least one school day in the previous year vs 39 % of the adolescents without pain. However, there were no differences in significant school loss (6 days or more) between these two groups. Robinson, Alvarez, and Dodge (1990) reported in a study of 40 referred children with RAP, that children with RAP had four times as many school absences as a matched normal control group. In a study by Walker and Green (1989), 41 children with RAP scored much higher than control children on the Functional Disability Inventory, which measures difficulties of functioning in the domains of home, school, recreation, and social interaction. The scores of the children with RAP were comparable to those with organic causes for their abdominal pain. Walker et al. (1995) showed in their follow-up study of 31 children with RAP that these children not only had persistent abdominal pain 5 to 6 years after diagnosis, but also continued to have more functional limitations and twice as many absences from school or work than controls. These results indicate that children with RAP suffer from pain and from limitations in their physical functioning but the question which aspects of physical functioning are most affected by RAP needs further clarification.

2.2.2. Psychological and social functioning

The psychological functioning of children with RAP has been the focus of many studies since the seminal study by Apley (1959). Apley found a clear relation between emotional disturbance and non-organic abdominal pain. Almost two-thirds of his series of children with RAP showed signs of emotional problems. Later studies approached this problem in a more specific way. Raymer, Weininger, and Hamilton (1984) found that the mean self-esteem scores of a group 16 children with non-organic abdominal pain was lower than those of 30 controls. The mean depression score of children with RAP was almost twice as high as that of the control children. Faull and Nicol (1986) found more psychiatric disturbance using the Rutter A(2) questionnaire in an epidemiological study among the population of 6-year-olds with RAP in a new town. Of the 48 cases with RAP, 31 showed clear signs of psychiatric disturbance. In a study among 13 children with RAP using a standardized child psychiatric interview and several standardized questionnaires, all children met criteria for a psychiatric diagnosis (Garber, Zeman, & Walker, 1990). These were mainly internalizing disorders including depressive disorders and anxiety disorders. In this study, the children with RAP obtained scores on the internalizing subscale of the Child Behavior Checklist (CBCL) comparable to scores of psychiatrically referred children.

The scores on the externalizing subscale of the CBCL of the children with RAP were not higher than those of normal controls. Walker and Green (1989) investigated a group of 41 children with RAP and compared these with 28 children with organic abdominal pain and 41 controls. They found that children with RAP scored higher on anxiety and depression self-report questionnaires than control children, but that they did not differ in these respects from children with organic abdominal pain. The mothers of the children with RAP, however, reported more internalizing problems on the CBCL for their children than mothers of children with organic abdominal pain and controls. In the follow-up study of these children, it was found that a large proportion of children with RAP continued to have higher levels of emotional problems than controls 5 to 6 years after diagnosis (Walker et al., 1995). In the study by Van Der Meer (1993) only a few differences were found between the psychological functioning of RAP children compared to the general population. These results may be explained by the fact that the instruments used to assess psychological functioning in this study were mostly personality inventories used to investigate a possible etiological relation between RAP and psychological factors. Emotional problems such as depression and anxiety are not well detected by these instruments and may have been missed. Hyams et al. (1996) found more anxiety and depression using standardized questionnaires among a community based population of adolescents with recurrent abdominal pain.

Social functioning has been poorly studied in children with RAP. Apley (1959) reported that the level of school functioning of children with RAP was comparable to that of control children. Faull and Nicol (1986) reported that 6-year-old children with RAP tend to settle less likely in school and dislike school more frequently than control children.

Little is known of the parent-child relation and family functioning of children with RAP. Robinson et al. (1990) concluded that children with RAP were more dependent and needed more attention than children from non-RAP control groups. In their study of 13 children with RAP, Garber et al. (1990) found that the mothers of children with RAP showed more symptoms of anxiety than mothers of children with organic abdominal pain and normal controls.

It can be concluded that a large proportion of children with RAP have problems in psychological functioning mainly of internalizing nature and that these problems appear to be persistent over time, while organic pathology is minor or absent even over longer time periods. So, RAP should be regarded as a problem which can only be explained within a bio-psycho-social model. In this model physical factors together with psychological, and possibly social factors play an important role in causing and maintaining the abdominal pain.

2.2.3. QL research in RAP

Future QL research in RAP should be aimed at identifying those aspects of a child's functioning which are most affected by RAP. More attention should be paid to limitations in functioning which are caused by RAP. These may concern limitations in participating in physical activities,

in school attendance, and in social activities. Together with measures of pain severity and measures of psychological functioning they can yield more information on the QL of these patients, in a broader sense than has been done until now. Eventually this is aimed at identifying those children with RAP who are most limited in their physical, psychological, and social functioning and who might benefit from specified treatment strategies.

2.3. Inflammatory bowel diseases

Inflammatory bowel diseases (IBD) are characterized by chronic inflammation of the small and/or large intestines of unknown origin. IBD is a collective heading for two diseases, Crohn's disease (CD) and ulcerative colitis (UC). The estimated incidence of CD is between 4 to 6 per 100,000 with 25 % to 40 % of cases under the age of 20 years, and the incidence of UC is 3 to 15 per 100,000 with 20 % of cases beginning in childhood or adolescence (Herbst, 1992). So, a total of 1.6 to 5.4 children or adolescents per 100,000 present yearly in childhood or adolescence with IBD.

In CD inflammation of the intestines is limited to the small bowel in 30 % to 35 % of cases, to the large bowel in 10 % to 15 % of cases, and affecting the small and large bowel at the same time in 50 % to 60 % of cases (Hyams, 1996). Clinical features of CD are gastrointestinal as well as extra-intestinal. Abdominal pain and bloody diarrhea are presenting symptoms in the majority of affected children. Abdominal pain is often severe and disturbing the child's sleep. However, presentation may also be obscure with abdominal pain and depression as the only symptoms, so that the delay between onset of disease and diagnosis may be 2 years (Büller, 1997). Anorexia, nausea, and vomiting are common concomitant complaints. Growth retardation and late onset of puberty are frequent in CD probably caused by chronic malnutrition and chronic inflammation. Apart from these symptoms, there is a myriad of possible extra-intestinal complaints affecting almost every tract (Hyams, 1996). Medical treatment is primarily aimed at reducing the inflammation process with anti-inflammatory agents such as 5-aminosalicylate and corticosteroid. Especially corticosteroid have many side-effects including growth suppression, moon facies, and depression. Surgery, aimed at resection of the affected part of the intestine, is necessary in 50 % to 70 % of cases within 10 to 15 years after diagnosis (Hyams, 1996). Hyams, Grand, Colodny, Schuster, and Eraklis (1982) reported that in 42 % of cases there was a relapse of inflammation within 5 years after colectomy requiring a second resection. None of these patients had more than one recurrence (Hyams et al., 1982).

UC shares many aspects of CD but localization in UC is limited to the large intestines. Clinical features are stools mixed with blood and mucus and lower abdominal cramping most intense during defecation. UC in children and adolescents tends to run a more complicated course than in adults with more frequent involvement of the entire colon (Kirschner, 1996). Delayed growth and sexual maturation are less frequent in UC than in CD. Arthralgia and arthritis are the

most frequent extra-intestinal complaints of children with UC (Kirschner, 1996). Medical and surgical treatment of UC is similar to that of CD. Recurrence of colitis after resection is reported to be much less frequent, i.e. 0 % in the series of Hyams et al. (1982). In short, CD and UC are chronic conditions with severe and prolonged symptomatology affecting the gastrointestinal as well as other tracts necessitating intensive medical and surgical treatment with potentially harming side-effects.

2.3.1. Physical functioning

2.3.1.1. Adult populations

Several measures exist for the assessment of health status of adult IBD patients (see for reviews: Garret & Drossman, 1990; Drossman, 1996). These measures can be distinguished in disease activity measures and disease specific QL measures. Disease activity measures are completed by physicians and include patients' symptomatology, results of physical examinations, and laboratory measures. These measures are useful in clinical practice but have limited value in QL assessment. Disease activity appears to be weakly related to overall QL in IBD patients (Turnbull & Vallis, 1995). Several instruments have been developed to assess IBD related QL (Drossman, 1996). These measures are in most cases questionnaires to be completed by patients. Most of these measures include items regarding bowel symptoms, systemic symptoms, social and emotional functioning, and functional impairment. Based on these studies, it appears that in adults with IBD QL is generally good, that patients with CD have greater disease severity and more impaired health-related QL than patients with UC, and that impairment in the psychological and social dimensions are greater than in the physical dimension (Drossman, 1996). These instruments were all developed for adults and no comparable measures, neither as self-report nor as parent-report, are available yet for children with IBD.

2.3.1.2. Pediatric populations

The physical symptoms of IBD are well described, the four most frequent symptoms of children with CD as well as UC being diarrhea, rectal bleeding, abdominal pain, and weight loss (Langholz, Munkholm, Krasilnikoff, & Binder, 1997). Physical functioning in pediatric populations has been studied far less than in adults. Langholz et al. (1997) studied disease activity of IBD in a geographically derived cohort of children comparing CD and UC. They found that 74 % (UC) to 83 % (CD) had moderate to high activity. Functional impairment due to IBD symptoms was not described. Rabbett et al. (1996) assessed the QL of 16 children with CD using a questionnaire which was read to each child with questions concerning a wide range of topics from disease activity to emotional and social functioning, and views about the future. The most common symptoms were abdominal pain and lack of energy. Almost all children expressed concerns about the side-effects of medication. Although all children indicated that they were doing well at school, two-thirds thought that they would do better without CD. Nineteen percent

to 44 % of the children experienced limitations in participation in physical education lessons and sports. In a recent study on the development of a QL measure for children with IBD, Griffiths et al. (in press) identified several important aspects of physical functioning which may be of concern for these children. Bowel symptoms, being bothered by having to take medicines, worries about surgery, and concerns about height, specifically for children with CD, were important issues. It is clear that physical symptoms of IBD in children can lead to considerable limitations in functioning and together with consequences of treatment procedures may affect their psychological and social functioning.

2.3.2. Psychological and social functioning

CD and UC were considered as classical 'psychosomatic' diseases for which psychological factors were believed to be a main cause. Personality was assumed to play a major role in the onset and course of these diseases (see for example Prugh, 1951). Finch and Hess (1962) proposed that children with UC should be considered as suffering from 'severe psychopathology often close to psychosis'. The main methodological flaw of these studies was that they mostly concerned psychiatrically referred patients. It must be stressed that the presence of a psychiatric disturbance or of emotional problems alongside a somatic disorder is no evidence that that somatic disorder is caused by the psychiatric disturbances. The studies by Helzer (Helzer, Stillings, Chammas, Norland, & Alpers, 1982; Helzer, Chammas, Norland, Stilling, & Alpers, 1984) did much to refute this psychosomatic hypothesis. They compared consecutive series of patients with CD and UC with control subjects with chronic diseases. UC patients did not have a higher frequency of psychiatric disturbances, while CD patients did show more obsessive-compulsive disorder and more depression (50 %) than controls. In these studies there was no evidence that IBD was consistently preceded or caused by a psychiatric disorder.

Although the cause of CD and UC is unknown, there is consensus over the fact that both diseases are immunologically mediated reactions triggered in a genetically determined susceptible host (Herbst, 1992). In this light, IBD should be regarded as chronic somatic diseases with possible negative consequences for psychological and social functioning.

Several recent studies have assessed the psychological and social functioning of children with IBD. In these studies two approaches are taken. The first is to investigate the presence of psychiatric disorders using semistructured psychiatric interviews (Burke et al., 1989a, 1989b; Engström & Lindquist, 1991; Steinhausen & Kies, 1982). The second method is to assess the amount of behavioral and emotional problems using standardized parent and child questionnaires (Engström, 1992; Steinhausen & Kies, 1982; Wood et al., 1987). An advantage of the last method is that the outcome of IBD children can be compared with normative data.

Steinhausen and Kies (1982) investigated a group of 17 children with IBD using a structured psychiatric interview and a behavior questionnaire. They found more than three times as many (60 %) psychiatric disorders, predominantly emotional disorders, among children with

IBD compared to a normal control group (18 %). Burke et al. (1989a, and 1989b) studied emotional problems in a consecutive series of children with IBD compared to children with cystic fibrosis (CF). The lifetime prevalence of depression was highest for CD compared to UC and CF, while children with UC had a higher lifetime prevalence for dysthymia and phobic disorder than CD and CF. The current prevalence of depressive disorders was higher in children with CD and UC than in children with CF. The highest rates for phobias, separation anxiety, and obsessive compulsive symptoms were found in UC. Engström and Lindquist (1991) found that psychiatric disorder, assessed using a reliable and well validated psychiatric interview, was four times as frequent in children with IBD (60 %) compared to a group of healthy children (15 %).

Steinhausen and Kies (1982) using the Children's Behavior Questionnaire found that children with IBD had more emotional problems but not more behavioral problems than normal controls. Wood et al. (1987) compared problem behavior of a group of children with CD, UC, and their siblings with normative data. They found that children with IBD obtained significantly higher total problem scores and higher internalizing problem scores on the CBCL than their siblings and than children in the norm groups. Engström (1992) investigated mental health and psychological functioning of a group of 20 children with IBD and compared them with children with chronic headache, diabetes mellitus, and healthy children. He assessed their functioning with a wide range of standardized instruments. Children with IBD and headache showed higher rates of total problems and internalizing problems on the CBCL than healthy children. Social competence of children with IBD was lowest among the three illness groups. According to the mothers, general well-being was worst for the children with headache, and according to their self-reports it was worst for the children with IBD. Low self-esteem and depression were more frequent for children with IBD and chronic headache than for children with diabetes or healthy children. It appeared that many children with IBD denied their emotional problems.

There are few data available relating psychosocial functioning to disease parameters. Wood et al. (1987) did not find a relation between disease severity and amount of problem behavior. This is comparable to the results of Tumbull and Vallis (1995) who did not find a relation between disease activity and QL in an adult population with IBD. So it is not clear if there are specific aspects of IBD, such as short stature in CD or side-effects of medication, which negatively influence psychosocial functioning. Very little is known about the effect of medical interventions on psychosocial functioning. Most studies concerned the effects of medical and surgical procedures on morbidity only. Lask, Jenkins, Nabarro, and Booth (1987) compared different aspects of functioning of 12 children with a stoma with 13 children who had been operated but had no stoma and 13 children who were treated medically. They found no differences in QL, psychosocial functioning, and self-esteem between these groups of children.

Family functioning of children with IBD has been poorly studied. Only one study presents some reliable data in this respect. Engström (1991a) compared family functioning of 20 children with IBD with healthy controls and children with diabetes. He concluded that families with a child with IBD showed more dysfunction than the other families. However, he concludes that

these results should be interpreted with some caution because the validity of the questionnaire was not fully presented. He also found that parents, especially mothers, of children with IBD experienced more distress and less social support compared to parents of a group of normal controls (Engström, 1991b). No relations were detected between these social parameters and disease activity. However, mothers with low social support, reported higher degrees of problem behavior for their children.

It may be concluded that QL in children with IBD is clearly reduced in the physical as well as in the psychological and social functioning domains. Physical symptoms are often serious and embarrassing, and side-effects of treatment are often a matter of concern. Limitations in social functioning due to the disease is considerable and emotional problems are frequent. Most studies do not find a correlation between objective measures of disease activity and QL. Although the ancient 'psychosomatic' hypothesis of IBD has been refuted, it has become clear that there is a complicated interplay between IBD, its effects on psychosocial functioning and relations within the family. Many aspects are still unclear and need further investigation. For example the fact that children with IBD show predominantly emotional problems and few behavioral disorders is still insufficiently explained.

2.3.3. QL research in IBD

In the future efforts should be made to develop QL measures for children and adolescents with IBD. Next to more objective measures of disease activity it will be necessary to include measures taking into account the subjective appraisal of disease severity and of the burden of treatment by the child. It will be necessary to assess those domains of functioning (school, peers, sports) in which a child is mostly limited by the disease or its treatment. The effort done by Griffiths et al. (in press) to adapt the IBD QL questionnaire for child and adolescent populations seems to be an important step in this direction. However, to these disease specific measures, generic measures of psychological and social functioning must be added. It will be necessary to study the QL of children with IBD not only in a cross-sectional design, but also to perform prospective longitudinal studies. In this way it will be possible to make a more funded balance of the positive and negative consequences of the disease and its treatment, and physicians, patients, and parents can be guided in the choice of treatment strategies.

2.4. Congenital abdominal anomalies

Congenital abdominal anomalies are in most cases life-threatening anomalies necessitating acute and sometimes lengthy hospitalization of the newborn and intensive surgical and medical treatment. As a consequence, the early physical development of children with these conditions is problematic. However, the impact of congenital abdominal disorders is not limited to the

neonatal period. Prolonged morbidity may negatively influence future QL of these children. The overall incidence of congenital abdominal anomalies can be estimated at 1 to 1.5 per 1000 live births. As such they are rare disorders but they form a large proportion of children in neonatal surgical intensive care units. Because congenital abdominal conditions are very different in initial presentation and subsequent physical problems, and because research on QL in these conditions differs considerably they will be treated separately in this chapter.

2.4.1. Congenital Diaphragmatic Hernia

Congenital Diaphragmatic Hernia (CDH) is characterized by a defect in the diaphragm leading to herniation of the gut into the pleural cavity compressing the lung. Also a hypoplasia of the lung on the contralateral side is seen, and it is likely that a primary abnormality in the developing lungs exists. The infant with CDH has respiratory difficulties immediately after birth and begins to swallow air. Consequently, the gastrointestinal tract is gradually filled with air compressing the lungs, which further worsens the respiratory functioning. The condition of the infant deteriorates rapidly and immediate hospitalization in a specialized center is necessary. Before ten to fifteen years ago the infant with CDH was operated immediately to repair the defect in the diaphragm, but nowadays management has changed to delayed surgery. The infant is first hospitalized in a surgical intensive care unit and operation is delayed until the condition of the child has been stabilized. Despite several developments in treatment procedures such as artificial ventilation and vaso-active medication, the mortality of CDH is still high. Only between 35 % and 45 % of children with CDH survive (Langham et al., 1996). The incidence of CDH is estimated between 0.24 to 0.36 per 1000 live births (Langham et al., 1996). Some 20 years ago Extra Corporeal Membrane Oxygenation (ECMO) was introduced in the treatment of CDH. ECMO is an intensive and costly treatment procedure for infants with respiratory distress. Although at first the outlook of survival for children with CDH treated with ECMO seemed more favorable this has recently been questioned (Langham et al., 1996).

2.4.1.1. Physical functioning

CDH is a disorder which primarily affects pulmonary and gastrointestinal functioning. Several authors report some residual defects in pulmonary functioning in children and adolescents with CDH (Chatrath, El Shafie, & Jones, 1971; Delepouille et al., 1991; IJsselstijn, Tibboel, Hop, Molenaar & De Jongste, 1997; Reid & Hutcherson, 1976; Wohl et al., 1977). In other studies no reduction in lung functioning is reported (Falconer, Brown, Helms, Gordon, & Baron, 1990; Freyschuss, Lännergren, & Frenken, 1984; Kerr, 1977; Wischermann, Holschneider, & Hubner, 1995). Although these reports seem contradictory, the overall view is that long-term pulmonary dysfunction of children with CDH during childhood is mostly minor and does not lead to severe functional impairment. Pulmonary dysfunction may become more pronounced in adulthood. Vanamo, Rintala et al. (1996) described ventilatory impairment in half of the adult survivors of

CDH. This outcome may be related to chest-wall deformities and scoliosis which are common among adults with CDH (Vanamo, Peltonen et al., 1996). Gastrointestinal symptoms are described by several authors. Especially gastroesophageal reflux with symptoms such as heartburn and regurgitation is common in patients with CDH (Kieffer et al., 1995; Vanamo, Rintala, Lindahl, & Louhimo, 1996). Although these symptoms are frequent, they are seldom disabling. So, despite the fact that CDH is a very serious condition with extensive physical problems in the newborn period long-term morbidity and physical limitations are limited.

2.4.1.2. Psychological and social functioning

The literature on long-term psychological and social functioning of children with CDH is very scarce. With the introduction of ECMO, which has great costs and was expected to enhance survival of children which have a very poor outcome, more attention has been paid to follow-up. Several studies have shown that more children than expected with CDH treated with ECMO had light to moderate cognitive developmental delays (D'Agostino et al., 1995; Lund et al., 1994; Stolar, Crisafi, & Driscoll, 1995; Van Meurs et al., 1993). These studies mainly concerned young to very young children, and longer follow-up periods are needed to appraise the significance of these findings. Only two studies reported on the cognitive development of children with CDH not treated with ECMO (Davenport, Rivlin, D'Souza, & Bianchi, 1992; Nobuhara, Lund, Mitchell, Kharasch, & Wilson, 1996). Davenport et al. (1992) reported that none of 23 children with CDH aged 18 to 94 months, not treated with ECMO but with delayed surgery, had developmental delays on the Griffiths' mental developmental scales. Based on the results of non-specified developmental examinations Nobuhara et al. (1996) concluded that children with CDH treated with ECMO had much more often developmental delays than CDH children not treated with ECMO. In a recent follow-up study of 11 children with CDH, Bouman, Koot, Tibboel, and Hazebroek (in press) found that the mean IQ of these children was 15 points below the norm of 100, indicating a significant delay in cognitive functioning. None of these children had been treated with ECMO.

In the same study, Bouman, Koot, Tibboel, et al. (in press) found that children with CDH, showed more emotional and behavioral problems as reported by parents and teachers, and more depressive problems as reported by the children themselves, compared to normative samples. No other follow-up studies on the psychosocial functioning of children with CDH are available. However, the conclusion seems justified that children with CDH, not only those treated with ECMO, are at risk for cognitive as well as psychosocial problems and that further studies will be necessary to evaluate these aspects of the QL of children with CDH.

2.4.2. Esophageal Atresia

Esophageal atresia (EA) is a congenital malformation of the esophagus which can have several forms. Incidence for all forms of atresia is estimated at 0.22 per 1000 live births. In the most

common form (85 % of the cases), the proximal part of the esophagus ends as a blind sac while the distal part is as a fistula connected to the trachea. In other cases there is only an interruption between the two parts of the esophagus. The earliest sign of EA is regurgitation of saliva. The first feeding is followed by choking, coughing, and regurgitation. Diagnosis is mostly made during the first days of life. Operation can be immediate or delayed depending on the type of anomaly, the condition of the infant, and the presence of other serious congenital anomalies. Due to narrowing of the constructed esophagus frequent dilatations under general anaesthesia may be required until 2 to 3 years of age. Half of the infants born with EA have other congenital anomalies. Especially cardiovascular, urogenital, and anorectal anomalies are common. Mortality rate varies from 0 % for children with normal birth-weights and absence of pneumonia to 43 % for children with low birth-weight, severe pneumonia, or other serious congenital anomalies. Overall survival is about 83 % (Ein, Shandling, Wesson, & Filler).

2.4.2.1. *Physical functioning.*

Feeding problems are common among patients with EA. Because the constructed esophagus never functions normally mainly due to the absence of peristaltic movements, patients with EA frequently complain of delayed passage of foods, the food being stuck in the esophagus with complaints of breathlessness, and heartburn due to reflux esophagitis (Anderson, Noblett, Belsey, & Randolph, 1992; Biller, Allen, Schuster, Treves, & Winter, 1987; Chetcuti & Phelan, 1993; Lindahl, Rintala, & Sariola, 1993; Puntis, Ritson, Holden, & Buick, 1990; Robertson, Mobaieek, Davis, & Coates, 1995; Saeki, Tsuchida, Ogata, Nakano, & Akiyama, 1988; Ure et al., 1994). In early childhood up to one-third of parents report severe feeding problems, but these tend to ameliorate in later childhood (Puntis et al., 1990). Although feeding problems persist into adulthood, they are seldom severe (Biller et al., 1987; Chetcuti & Phelan, 1993; Ure et al., 1994). Pulmonary problems, consisting of lower airway disease, increased respiratory symptoms, and reduced pulmonary function are reported in up to 40 % of patients (Chetcuti, Phelan, & Greenwood, 1992; Robertson et al., 1995). Overall, 90 % of patients with EA have good to excellent outcome with no or minor gastrointestinal or pulmonary dysfunction. Between 19 % and 35 % of patients with EA have skeletal anomalies of the spine and thorax (Chetcuti, Dickens, & Phelan, 1989; Chetcuti, Myers, Phelan, Beasley, & Dickens, 1989) and these may contribute to the pulmonary problems, which may become more pronounced with increasing age as is the case in CDH. Children with EA may be small and relatively light probably due to prolonged feeding problems. Growth retardation is described by Ahmed & Spitz (1986), Anderson et al., 1992, and Puntis et al., (1990).

So, EA is a condition causing long-term gastro-intestinal and pulmonary morbidity with, however, limited functional impairment in about 10 % of cases.

2.4.2.2. *Psychological and social functioning*

Very few studies concerned the psychological and social consequences of EA. The earliest study is by Dera, Mies, and Martinus (1980), who assessed the psychosocial functioning of 10 children with EA with a mean age of 4.3 years. They found a mean IQ of almost 10 points below the norm of 100 and frequent symptoms of anxiety, regression, and disturbances of contact. Lehner (1989) sent a questionnaire to parents of children with EA who were member of a patient organization for congenital abdominal anomalies. One hundred and twenty-two parents responded with children aged 0 to 19 years. They reported that the mental and physical development of most patients was normal and that most children visited normal schools. No further specifications were given by the author. Chetcuti, Myers, Phelan, and Beasley (1988) followed 125 children with EA into adulthood. They reported that almost all enjoyed a normal lifestyle. Employment and marital status were in the normal range. Ure et al. (1994) investigated the outcome in adulthood of 8 patients with EA after colon interposition. Two patients were slightly mentally disabled. Seven patients reported unimpaired QL on the Spitzer index. Bouman, Koot, and Hazebroek (in press) studied the psychological and social functioning of 36 children with EA using interviews and standardized assessment procedures. The mean IQ of these children was almost 10 points lower than the norm of 100. Children with major associated congenital anomalies who had been artificially ventilated in the newborn period (n' 8) even had a mean IQ more than 20 points below the norm. More than twice as many children (30 to 35 %) as in the general population (15 %) showed elevated rates of emotional and behavioral problems as reported by parents and teachers. The children themselves did not report more negative self-esteem or more depressive symptoms than children in the general population.

EA appears to have only slight negative consequences for adults, but younger children with EA seem to suffer more from emotional and developmental difficulties. These may be related to the long period of separation from the parents when they were infants and the frequent feeding difficulties (Dera et al, 1980) or to other risk-factors such as the presence of other associated congenital anomalies and artificial ventilation in the newborn period (Bouman, Koot, & Hazebroek, in press).

2.4.3. **Abdominal Wall Defects**

Children with abdominal wall defects (AWD) are born with a large abdominal mass protruding through a defect of the abdominal wall without (gastroschisis) or with (omphalocele) a covering amniotic membrane. Mortality was high before the introduction of total parenteral nutrition. Operation is performed after stabilizing the infant. Mortality is nowadays between 10 and 20 % (Halsband & Von Schwabe, 1989).

2.4.3.1. *Physical functioning*

The earliest report on the long-term functioning of children with AWD is from Touloukian and Spackman (1971). They found normal growth in all and normal gastrointestinal function in five of six patients. Generally good health, few gastrointestinal problems, and the absence of physical limitations were reported for children and adolescents with AWD (Daum, 1984; Halsband & Von Schwabe, 1989; Larsson & Kullendorf, 1990; Lindham, 1984; Swartz, Harrison, Campbell, & Campbell, 1986; Tunell, Puffinbarger, Tuggle, Taylor, & Mantor, 1995). The absence of a navel or a disfiguring abdominal scar was reported to be a point of embarrassment for 20 to 25 % of the patients, especially for girls (Lindham, 1984; Halsband & Von Schwabe, 1989; Tunell et al., 1995).

2.4.3.2. *Psychological and social functioning.*

Very few data are available on the psychological and social aspects of the QL of children with AWD. Swartz et al. (1986) reported satisfactory academic performance for 25 school-aged children. In a follow-up study by Tarnowski, King, Green, and Ginn-Pease (1991) including 22 children with AWD, intelligence, reading, and mathematical abilities were tested. The mean scores on these academic measures were all within normal limits. The same study assessed the level of problem behavior and social competence using the CBCL. The children with AWD showed more emotional and behavioral problems than could be expected based on the normative data. About 18 % of the children obtained total problem scores in the clinical range (versus the norm of 10 %) and they showed as much emotional problems (28 % internalizing scores in the clinical range) as behavioral problems (25 % externalizing scores in the clinical range) (Tarnowski et al., 1991). The children with AWD obtained lower social competence scores than in the norm group (23 % in the clinical range). In the same study children with anorectal anomalies (ARM) were included. No differences were found between children with AWD or with ARM on any of the outcome measures. This is counterintuitive because children with ARM were expected to have worse psychological functioning caused by their life long physical problems especially incontinence, while children with AWD are relatively free of physical problems after the neonatal period. It can be hypothesized that the embarrassment about the absence of a navel and the abdominal scar, which are both clearly visible defects, may have a negative influence on self-esteem and hence contribute to behavioral problems. So, there is reason to perform further studies on the QL of children with AWD.

2.4.4. **Intestinal atresia***

Intestinal atresia (IA) can be localised in the duodenum, jejunum, or ileum. Estimated incidence of duodenal atresia is much lower (1 in 20,000 to 40,000 births; Schnauffer, 1986) than of jejunoileal atresia (1 in 330 to 1,500 births; Grosfeld, 1986). Atresia refers to a congenital

* This section was added to the original chapter text.

obstruction of the intestinal lumen due to complete occlusion (95 % of cases) or stenosis. The clinical presentation consists of the onset of bilious vomiting within a few hours after birth, abdominal distention, jaundice, and failure to pass meconium in the first days of life. Treatment consists of resection of the obstructed part of the small bowel, after stabilisation of the infant. The resection of a part of the small bowel results in the so-called 'short bowel syndrome' characterized by intestinal malabsorption necessitating long-term parenteral nutrition. In most cases a normal feeding and defecation pattern and normal growth is reached within the first three years of life (Brand et al., 1989). The prognosis of IA has been much improved due to the introduction of parenteral nutrition, and improvement of operation and life-supporting techniques leading to current survival rates between 80 % and 95 % (Goulet et al., 1991; Grosfeld & Rescorla, 1993). Associated congenital anomalies are frequent in IA. The most frequent associated anomalies are abdominal and cardiac anomalies, and in up to one-third of cases IA is associated with Down syndrome (Grosfeld & Rescorla, 1993).

2.4.4.1. Physical functioning

A few studies have assessed some aspects of physical functioning of children with IA. Brand et al. (1989) investigated 16 children aged 2 to 12 years with short bowel syndrome, half of whom had IA. The mean body length of the children was on the 20th percentile. The physical functioning of the children was good with only slight limitations in activities due to an increased sensitivity to diarrhea during upper respiratory tract infections in half of the children. Affourtit, Tibboel, Hart, Hazebroek, and Molenaar (1989) investigated the long-term consequences of bowel resection in the neonatal phase of life in a group of 59 children aged 4 months to 11 years, of whom 25 % had IA. They found normal growth in most cases. The long-term physical functioning was rated as good with mostly normal defecation patterns and slight fat intolerance in some cases. Finally, Kokkonen, Kalima, Jääskeläinen, and Louhimo (1988) assessed the late follow-up of 41 adult patients aged 15 to 35 years with duodenal atresia. Twenty-eight patients were completely free of symptoms, 10 reported some discomfort due to mild gastro-intestinal symptoms, and only 3 had major gastro-intestinal pain.

It can be concluded that the physical functioning of patients with intestinal atresia is generally good with mild gastro-intestinal symptoms in a minority of cases.

2.4.4.2. Psychological and social functioning

In the one follow-up study concerning psychosocial functioning of patients with IA, Brand et al. (1989) did not find increased behavioral or emotional problems in 16 children with short bowel syndrome compared to normative data. No studies are performed on the family functioning of children with IA. Obviously, no conclusions can be drawn concerning the psychological or social functioning of children with IA based on these scarce empirical data.

2.4.5. Hirschsprung's Disease and anorectal malformations

Although Hirschsprung's disease (HD) and anorectal malformations (ARM) are different anomalies they share many characteristics especially concerning long-term functioning. This is why they will be described together. The nature of HD is the absence of ganglion cells in the distal intestine. This aganglionosis not only can involve the lower rectum, but can extend higher into the colon and even into the small intestine. The absence of ganglion cells causes abnormalities of the intestinal peristaltic movements and accounts for a functional obstruction. This obstruction causes a widening, megacolon, proximal to the aganglionic segment. The presenting symptomatology is variable from complete obstruction at birth with vomiting and abdominal distention to a long-standing mild constipation. Final diagnosis is made by radiological examinations, a rectal biopsy, and rectal manometry, but may be delayed several years in some cases due to mild symptomatology. Treatment consists of resecting the aganglionic bowel and connecting normal bowel to the distal rectum. The incidence of HD is about 1 in 5000 live births. Mortality is around 5 % mainly due to ischemic enterocolitis in the neonatal period (Rescorla, Morrison, Engles, West, & Grosfeld, 1992).

In ARM, the anus ends in a blind loop with often a fistula between the blind anal canal and the urethra in men and the genital region in women. In some cases there is an anus visible from the outside and in other cases the normal anal opening is covered with skin. Not only does the anus end in a blind loop but also the innervation and musculature of the pelvis and anorectum are partly or completely absent. Depending on the anatomical level of the anomaly ARM is divided in high and low ARM. Due to its localization high ARM is surgically the most complicated. Presenting symptoms of ARM are the absence of an anus or signs of fistulation on inspection of the neonate and absence or abnormal passage of meconium, the infants first stools. Treatment is surgical in the vast majority of cases. In some cases when there is only a narrowing of the anal canal progressive dilatation will be sufficient. In low ARM, the surgical procedure is less complicated. However, after repair daily dilatations of the anus are necessary for several months to a year. In high ARM, surgical procedures are often very difficult due to the localization and the concomitant vesico-urinary anomalies. A staged procedure with a colostomy is sometimes necessary. Incidence of ARM is around 1 in 5000 live births. ARM often is associated with other serious congenital anomalies such as genito-urinary, cardiovascular, or central nervous system anomalies, which may be the primary cause of death.

2.4.5.1. Physical functioning

The most problematic consequences of HD and ARM are continence related problems. Defecation appears to be a very complex process in which anatomical, physiological, and neurological factors are finely tuned in order to obtain such a simple thing as fecal continence.

Due to congenital anomalies or surgical procedures this process is disrupted leading to a high frequency of continence problems or constipation in HD and ARM. Continence reported in HD is about 65 % in most recent studies (Heij, De Vries, Bremer, Ekkelkamp, & Vos, 1995; Marty et al., 1995; Moore, Albertyn, & Cywes, 1996; Rescorla et al., 1992). In one study only 20 % of patients obtained normal continence (Heij et al., 1995), a discrepancy which cannot be explained by the authors on the basis of patient selection or follow-up method. The figures for ARM are worse. Normal continence, is reached by maximally 60 % of patients in one study (Rintala, Mildh, & Lindahl, 1992), and of these only 15 % had optimal continence (i.e. no signs whatsoever of smearing of feces in underwear or constipation). Rintala et al.(1992) were the only authors who compared continence of ARM patients with normal controls. In normal controls 100 % had normal continence and 76 % optimal continence. Younger children appear to have the greatest problems as is shown in a study by Langemeijer and Molenaar (1991) who found that none of 50 children aged 4 to 7 years were continent, although 40 % reached pseudo continence with daily enemas. In a study of 50 children with HD or ARM, Bouman, Koot Langemeijer, and Hazebroek (submitted) found that almost one-third of children with ARM had moderate to serious fecal incontinence versus none of the children with HD. Diseth and Emblem (1996) reported that of 33 adolescents with ARM 77 % had impaired control of continence with 40 % having occasional staining and 37 % having intermittent or constant soiling. The figures for adults are somewhat better as is shown in the study of Rintala et al. (1992) and Hassink, Rieu, Brugman, and Festen (1994). Although Hassink et al. (1994) reported that none of 56 adults reached complete continence, 84 % of them had 'socially acceptable' continence. An age effect is also shown by Ditesheim and Templeton (1987). Continence increased with increasing age, with 33 % of children aged 2.5 to 9 years, 58 % of adolescents aged 10 to 16 years, and 63 % of adults aged 17 to 24 years having normal continence. Apart from gastrointestinal problems, urologic problems are reported in 24 to 29 % of patients with ARM (Boemers, De Jong, Van Gool, and Bax, 1996; Misra, Mushtaq, Drake, Kiely, and Spitz, 1996). These problems concern incontinence, vesico-ureteric reflux, recurrent urinary tract infections, and eventually renal failure. However, concerns of parents about fecal incontinence appear to be much greater than about urinary incontinence and urologic problems (Boemers et al., 1996).

2.4.5.2. Psychological and social functioning

Psychological and social functioning of HD and ARM are the most extensively studied among congenital abdominal anomalies. As in IBD, a difference must be made between studies assessing the prevalence of psychiatric disorders and studies assessing the amount of emotional or behavioral problems in children with HD or ARM. Using the Child Assessment Schedule (CAS) Ludman, Spitz, and Kiely (1994) found that 29 % of 160 children with ARM aged 6 to 17 years had a mild to moderate psychiatric disorder most of which were internalizing disorders. Diseth

and Emblem (1996) even found that 58 % of 33 adolescents with ARM had a psychiatric disorder, also predominantly internalizing. In 19 adolescents with HD Diseth et al. (1997) did not find increased rates of psychiatric disorder.

Several studies have assessed the rates of behavioral and emotional problems using the CBCL and related questionnaires (Teachers Report Form [TRF], Youth Self report [YSR]). The outcomes of these studies are contradictory. Diseth found no increased behavioral and emotional problems in adolescents with ARM (Diseth and Emblem, 1996) and with HD (Diseth et al., 1997). There is a contradiction in the fact that in the study of adolescents with ARM, Diseth and Emblem (1996) found 58 % of them having psychiatric disorder, while CBCL and YSR total problems scores were in the normal range. The explanation by the authors that internalizing problems are often underreported seems insufficient and it raises questions about the methodology used in the study. Higher rates of emotional and behavioral problems were found in studies by Ludman et al. (1994) concerning 160 children and adolescents with ARM, by Ginn-Pease et al. (1991) concerning 34 children with ARM, and by Bouman, Koot, Langemeijer, et al. (submitted) concerning 50 children with ARM or HD. Until now there are no indications that children with HD or ARM have lower self-esteem. In the studies by Ginn-Pease et al. (1991) and Bouman, Koot, Langemeijer, et al. (submitted) mean scores on respectively the Piers-Harris Self-Concept Scale and the Self-Perception Profile for Children were within normal limits. There are however indications that age plays a role in the development of psychosocial problems in children with HD or ARM. Adolescents with ARM seem to be at greater risk for emotional and behavioral problems (Diseth & Emblem, 1996) or negative self-concept (Ginn-Pease et al., 1991). Because fecal incontinence is supposed to have the most negative influence on psychosocial functioning of patients with ARM or HD, most studies have assessed the relationship between fecal incontinence and psychosocial outcome measures. Ginn-Pease et al. (1991) found increased rates of maladjustment for children who were incontinent compared to continent children. Other authors however did not find increased psychiatric disorders or behavioral and emotional problems for incontinent children compared to continent children (Ludman et al., 1994; Diseth & Emblem, 1996; Diseth et al., 1997; Bouman, Koot, Langemeijer, et al., 1998c). However, Diseth found a negative influence of incontinence on overall psychosocial functioning using the Children's Global Assessment Scale (CGAS) (Diseth & Emblem, 1996; Diseth et al., 1997). No negative influence of incontinence on self-concept of children with ARM was found by Ginn-Pease et al. (1991) and Bouman, Koot, Langemeijer, et al. (submitted).

Social problems related to continence are reported for 12 % to 39 % of adults with ARM (Hassink et al., 1994; Rintala et al., 1992). Ditesheim and Templeton (1987) developed a QL measure focusing on the social limitations due to continence problems. Items included in the measure were school attendance, limitations in social relations, and restrictions in physical activities. Based on this measure the QL could be rated as good, fair, or poor. Seventy-two

percent had good, 23 % fair, and 5 % poor QL. However, this conclusion is not relevant to QL in general, because in this study, QL was directly related to the effects of continence problems on social functioning.

Diseth and Emblem (1996) found that the duration of anal dilation in children with ARM in early childhood was negatively correlated to mental health and psychosocial functioning. No other studies have approached this problem, but it is an important finding because it indicates that frequent painful medical procedures in early childhood may have a long-standing negative influence on the psychological development of a child.

It can be concluded that HD and ARM are congenital anomalies which have long-term negative consequences for physical, psychological, and social functioning. Fecal incontinence is an embarrassing problem which affects a large proportion of children with HD or ARM. Many children with HD and ARM who have continence problems are functioning very well. However, in adolescence incontinence may have a more negative influence on QL. It appears that incontinence is not the sole determinant of QL in these children. Other factors, such as having a congenital anomaly regardless of its consequences, or undergoing frequent painful medical procedures, may be of equal importance for the QL of these children. This is supported by the results of a large follow-up study of 139 children with several congenital abdominal disorders (Bouman, Koot, Verhulst, & Van Gils, in press). In this study the children with congenital abdominal anomalies did function worse than normative groups in several aspects (intelligence, school level, problem behavior) but very few specific aspects of the different anomalies influenced QL.

2.4.6. QL research in congenital abdominal anomalies

In conclusion, it can be said that there are ample indications that it is necessary to continue QL research in children with congenital abdominal anomalies. The most adequate strategy seems to be to use generic QL measurers which cover in a comprehensive way the physical, psychological, and social domains. Only for some anomalies specific measures will have to be added, such as measures for fecal continence in HD and ARM. In addition, more attention should be paid to the impact on the family functioning of children with congenital abdominal anomalies, but of children with RAP and IBD as well, because this is a very poorly studied subject in these populations. More efforts should be made to identify factors that determine QL. Application of QL measures might be used in regular follow-up procedures for these children which may help to implement adequate treatment and preventive measures.

Chapter 3

Outline of the study

3.1. Aims of the study

The goals of the present study were to assess in a reliable and valid way the physical, psychological, and social functioning and its determinants of children with congenital abdominal anomalies. To this end an unselected cohort of children with congenital abdominal anomalies was studied with a broad range of standardized assessment instruments concerning their physical and psychosocial functioning and concerning environmental variables such as family functioning and psychosocial stress.

3.2. Subjects

3.2.1. Study group

The sample consisted of 139 children, 87 boys and 52 girls, who were born in the period from 1982 to 1986 and were treated in a large university children's hospital for a major congenital abdominal anomaly. A total of 226 children were treated in the hospital in the indicated time-period, of which 166 children survived. Of these 166 surviving children, 115 children and the parents of 139 children consented to participate leaving a total response of 83.7 %. The mean age of the study group children at the time of the assessment was 10.6 years (SD 1.1). One hundred and nineteen children lived with both parents, 19 children of divorced parents lived with one parent, and 1 child was institutionalized. The distribution of socio-economic status (SES), based on the highest parental occupational level (Van Westerlaak, Kropman, & Collaris, 1975), was as follows: low SES n=25, middle SES n=62, and high SES n=52.

3.2.1.1. Medical characteristics

The sample consisted of children with six so-called index diagnoses which represent the most frequent major congenital abdominal anomalies: esophageal atresia, diaphragmatic hernia, abdominal wall defects, intestinal atresia, Hirschsprung's disease, and anorectal malformations. Almost all children were hospitalized in the first two days after birth, except the children with Hirschsprung's disease who were sometimes hospitalized several months after birth. The length of the first hospitalization, the number of subsequent hospital admissions and operations were considerable for many children. These data are summarized in Table 3.1. Many children had associated congenital anomalies. The frequency of these anomalies and their nature are given in Tables 3.2 and 3.3

Table 3.1
Medical characteristics of the study sample

Diagnosis	N	First Hospitalization		Operations	Hospitalizations
		Age days ¹	Duration days ²	to follow-up n ³	to follow-up n ⁴
Esophageal atresia (EA)	31	2* (1 - 2)†	54 (13 - 690)	4 (0 - 6)	2 (1 - 18)
Diaphragmatic hernia (DH)	11	1 (1 - 2)	33 (10 - 180)	1 (1 - 4)	4 (1 - 10)
Abdominal wall defect (AWD)	19	1 (1 - 2)	57 (6 - 300)	2 (1 - 5)	2 (1 - 8)
Intestinal atresia (IA)	26	2 (1 - 22)	45 (14 - 330)	2 (1 - 5)	1 (1 - 8)
Hirschsprung's disease (HD)	23	4 (1 - 1000)	26 (4 - 106)	3 (1 - 7)	4 (1 - 13)
Anorectal malformation (ARM)	22	1 (1 - 3)	17 (12 - 78)	3 (0 - 8)	5 (1 - 25)
Multiple diagnoses (MD)	7	1 (1)	90 (42 - 315)	4 (2 - 5)	5 (2 - 13)

* Median (Range).

Results of analyses of variance:

¹ HD > EA, DH, AWD, IA, ARM, MD (F 3.81, p < .01).

² MD > DH, IA, HD, ARM; EA > HD, ARM; AWD > ARM (F 2.81, p < .05).

³ MD > EA, DH, AWD, IA; ARM > EA, DH, AWD, IA; HD > DH, IA (F = 4.69, p < .001).

⁴ ARM > AWD, IA; HD > AWD, IA, MD; MD > IA; EA > IA (F = 3.29, p < .01).

Table 3.2

Associated congenital anomalies

Diagnosis	N	Associated congenital anomalies		
		None	Minor	Major*
Esophageal atresia (EA)	31	13	12†	6
Diaphragmatic hernia (DH)	11	5	4	2
Abdominal wall defect (AWD)	19	9	7	3
Intestinal atresia (IA)	26	21	1	4
Hirschsprung's disease (HD)	23	22	0	1
Anorectal malformations (ARM)	23	6	13	3
Multiple diagnoses (MD)‡	7	0	0	7

* Major associated congenital anomalies are life-threatening anomalies and any neurological or chromosomal anomaly. All other anomalies are classified as minor.

† One child may have more than one associated congenital anomaly.

‡ EA + ARM: n=3; EA + AWD: n=1; AWD + IA: n=1; AWD + ARM: n=1, EA + IA + ARM: n=1

Table 3.3

Nature of associated congenital anomalies

Minor:	Major:
Anomalies of trachea or larynx	Second major abdominal anomaly
Meckel's diverticulum	Ventricular septum defect
Skeletal anomalies	Tetralogy of Fallot
(including palatoschisis)	Chromosomal anomalies
Open ductus arteriosus	Hydrocephalus
Non-descended testes	Spina Bifida
Urogenital anomalies	
Renal agenesis of one kidney	
Microtia	
Annular pancreas	

3.2.2. Comparison group

A comparison group was enrolled, which was recruited from the general population. The comparison group consisted of 136 children, 83 boys and 53 girls, mean age 10.5 years. The comparison group was selected as follows. The municipal council of six villages and towns with degrees of urbanization corresponding to those of the study group children were approached and they gave names and addresses of a number of randomly selected children with an age and gender distribution corresponding to that of the study group. These children and their parents were approached by telephone to ask their participation and obtain their informed consent. The response rate of the comparison group is estimated to be 60 %. Due to the fact that the municipal councils used different procedures to approach the subjects it is not possible to be more specific

about response rate, neither to give information on reasons for refusal. Eventually, the comparison group matched with the study group on the variables gender, age, and degree of urbanization.

One hundred and thirty-one children in the comparison lived with both parents, and the parents of 5 children were divorced. One of these children lived with foster parents. No children in the comparison group were institutionalized. The distribution of SES was as follows: low SES n=23; middle SES n=54, and high SES n=59.

The comparison group was used in the development of the quality of life instrument (Chapter 4). In the rest of the study normative data were used to compare study group scores on standardized instruments.

3.3. Measures

The instruments used in this study are summarized in Table 3.4.

3.3.1. Sociodemographic variables

Information was obtained from the parents about living arrangements, marital status, socio-economic status, and type of school and grade.

3.3.2. Physical functioning

The children in the study group and one of their parents were seen in the hospital for a physical examination of the child performed by a pediatric surgeon.

3.3.3. Intelligence, school competence, and adaptive behavior

Intelligence was tested with two subtests, Vocabulary and Block Design, of the Wechsler Intelligence Scale for Children-Revised edition, which together have a correlation of .92 with full scale IQ (Van Haassen et al., 1986). Indicators of school competence were school level and the School Competence and Academic Performance scales of the Child Behavior Checklist and Teacher Report Form (further descriptions see below). Adaptive behavior in three domains, 'communication', 'daily living', and 'socialization' was assessed with the Vineland Adaptive Behavior Scales (VABS; Sparrow, Balla, & Cicchetti, 1984). Scale scores for each of these domains and a total score can be computed.

Table 3.4
Overview of instruments

Aspect of functioning	Instrument	Completed by			
		Child	Mother	Father	Teacher
<i>Child functioning</i>					
Physical functioning	Physical examination	112*	111	-	-
Intelligence	Wechsler Intelligence Scale for Children-Revised	115	-	-	-
Adaptive behavior	Vineland Adaptive Behavior Scales	-	112	-	-
Psychosocial functioning	Interview	111	114	-	-
Self-esteem	Self-Perception Profile for Children	105	-	-	-
Depression	Abbreviated Depression Questionnaire for Children	111	-	-	-
Health-related quality of life	Quality of life questionnaire for children	110	135	124	-
Problem behavior	Child Behavior Checklist Teacher's Report Form	-	134	112	-
		-	-	-	118
<i>Psychosocial stressors</i>					
Life events	Life-Events Questionnaire	-	137	122	-
Disease related stress	Interview	111	114	-	-
<i>Social-ecological variables</i>					
Family functioning	Family Assessment Device	-	136	120	-
Marital relationship	Interactional Problem Solving Questionnaire	-	121	108	-
Parental psychopathology	General Health Questionnaire	-	133	118	-

* Number of completed forms.

3.3.4. Health-related quality of life of the children

Health-related quality of life was assessed with the *Quality of Life Questionnaire for children* (Bouman, Koot, Van Gils & Verhulst, in press) which will be described in chapter 4. The quality of life questionnaire for children was completed by both children and parents.

3.3.5. Psychosocial functioning of the children

3.3.5.1. Self-reports

A 'psychosocial interview' of the children concerning general psychosocial functioning was performed. This interview contained items concerning the functioning of the child within the family, at school, and in relation to peers.

Self-esteem and depression were assessed with the *Self-Perception Profile for Children* (SPPC; Harter, 1985; Van Dongen, Koot, & Verhulst, 1993) and the *Abbreviated Depression Questionnaire for Children* (ADQC; De Wit, 1987).

3.3.5.2. *Parent reports*

Also with one of the parents a psychosocial interview was conducted containing the same questions as with the children.

Behavioral and emotional problems of the children were measured with the Child Behavior Checklist (CBCL, Achenbach, 1991a; Verhulst, Van Der Ende, & Koot, 1996). The CBCL includes a problem section containing 120 items concerning emotional and behavioral problems and a competence section reflecting school functioning and social competence of the children.

3.3.5.3. *Teacher-reports*

The teachers completed the Teacher's Report Form (TRF, Achenbach, 1991b; Verhulst, Van Der Ende, & Koot, 1997) The TRF is a teacher version of the CBCL. The problem part of the TRF is scored on the same scales as the CBCL. The competence scales of the TRF are Academic Performance and Total Competence comprising items such as attitudes toward learning, appropriate behavior, and happiness.

3.3.6. **Psychosocial stressors**

General psychosocial stress in the participating families was measured with the Life-Events Questionnaire (LEQ; Berden, Althaus, & Verhulst, 1990). In addition, disease related psychosocial stress was assessed by interviewing the parents and children on those problems in the three main domains of functioning, family, school, and peers, which were specifically related to the congenital anomalies.

3.3.7. **Social-ecological variables**

Family functioning was assessed with the General Functioning Scale of the Family Assessment Device (FAD; Kabacoff, Miller, Bishop, Epstein, & Keitner, 1990; Miller, Bishop, Epstein, & Keitner, 1985). As measure of the marital relationship the Interactional Problem Solving Questionnaire (IPSQ; Lange, 1983) was used. The IPSQ measures the problem solving abilities of partners in a marital relationship and can be considered as a unidimensional measure of the quality of the marital relationship. Psychopathology of the parents was measured with the 12-item version of the General Health Questionnaire (GHQ-12; Koeter, & Ormel, 1991). The GHQ-12 is a measure for non-psychotic psychological problems, especially emotional problems such as depression and anxiety.

3.4. Procedure

The research protocol was reviewed and approved by the medical ethical board of the hospital. The children in the study group and comparison group and their parents received a letter in which they were informed about the purpose and procedures of the study. They were contacted by telephone to ask their participation and obtain their informed consent.

The 115 participating children underwent a physical examination, were interviewed and subjected to an abbreviated intelligence test, and they completed the QLQC, SPPC, and ADQC and their parents were interviewed. The participating parents of 139 children completed the QLQC and CBCL at home. One-hundred-and-thirty-three children in the comparison group consented to participate and completed the QLQC, SPPC, and ACDQ in small groups in a school in the neighborhood where they lived. The parents of all 136 children in this group completed the same questionnaires as the parents of the study group children at home. Questionnaires were completed by both parents of 120 children in the study group and of 126 children in the comparison group and in all other cases only by the mother. The parents were instructed to complete the questionnaires independently.

Chapter 4

Development of a health-related quality of life instrument for children: The Quality of Life Questionnaire for Children.

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Psychology and Health (in press)

4.1. Abstract

Developed a generic self-report and parent-report health-related quality of life instrument: the Quality of Life Questionnaire for Children (QLQC). One hundred and thirty-nine 8- to 12- year old children with congenital gastro-intestinal anomalies and a matched comparison group of 136 children from the general population participated. The QLQC has 118 items, 13 scales, covering the physical, psychological, and social functioning domains of children. Cronbach's alphas of the scales ranged from .36 to .91 and test-retest correlations from .36 to .89. On the QLQC, study group children showed more physical problems ($p = .002$ to $.011$) and more cognitive problems ($p = .015$ to $.039$) than comparison group children. The QLQC discriminated well between children with and without emotional and behavioral problems. QLQC scales showed high correlations with comparable scales of concurrent instruments. It can be concluded that the psychometric properties of the QLQC are promising but that further research is needed.

4.2. Introduction

Chronic health conditions, including both chronic diseases and impairments caused by either congenital or acquired conditions, affect a large proportion of children. Based on the US National Health Interview in 1988, Newacheck and Taylor (1992) estimated that 31 % of children under 18 years of age were suffering of chronic health conditions. The impact of chronic health conditions in childhood concerns not only the children's physical but also their psychological and social functioning. These children have frequent physical complaints, such as pain, tiredness, or

breathlessness, and they are limited in their daily physical activities such as going to school or participating in sports (Stein & Jessop, 1990; Walker & Greene, 1991). Numerous studies have shown that children with chronic diseases are at risk for problems in psychosocial adjustment (Eiser, 1990; Lavigne & Faier Routman, 1992; Pless & Nolan, 1991; Wallander & Varni, 1998). Concerning congenital anomalies, emotional and behavioral problems are reported for children with congenital heart disease (Kramer, Awiszus, Sterzel, Van Halteren, & Claßen, 1989; Utens et al., 1993), craniofacial deformities (Pruzinsky, 1992), congenital orthopedic deformities (Kashani, Venzke, & Millar, 1981; Varni & Setoguchi, 1996), and hearing defects (Heller, Rafman, Zvagullis, & Pless, 1985). Those few studies that are concerned with the long-term functioning of children with a major congenital abdominal anomaly showed that these children are also at risk for problems in physical, psychological, and social functioning (Chetcuti & Phelan, 1993; Diseth & Emblem, 1996; Ginn-Pease et al., 1991; Kato et al., 1993; Vanamo, Rintala, Lindahl, & Louhimo, 1996).

These concerns with the functioning of children with chronic health conditions have led to increasing interest in the concept and measurement of quality of life of childhood physical conditions, usually referred to as health-related quality of life (HRQOL). Spieth and Harris (1996) discerned two main conceptual models of HRQOL, the utility model developed among others by Kaplan and Anderson (1988), and the health status measurement model. In the utility model values are assigned to different health states based on judgments by research experts, physicians, or lay people. These values can be used in cost-effectiveness analyses of medical treatments. (Torrance, 1986). Spieth and Harris (1996) argued that "the utility model may not be applicable to pediatric populations because children have difficulty understanding and formulating abstract preferences between quality and quantity of life". As a result the health status measurement approach has been adopted in the majority of HRQOL research in pediatric populations.

In 1947 the World Health Organization defined health as a state of complete physical, mental, and social well-being. Starting from this definition there is general agreement that HRQOL is a multidimensional construct including three broad domains, i.e. the physical, psychological, and social functioning domains (Aaronson, 1991; Kaplan, 1988; Spieth & Harris, 1996). However there is debate if this multidimensionality should be reflected in outcome measures. It is argued that it is very difficult to compare the HRQOL of different persons if their HRQOL is affected in different domains (Kaplan, 1988). A unidimensional approach giving an overall summary measure of quality of life would then be preferred. However, for children this approach raises several questions such as how to value the different domains of their life, by who this should be done, and how the developmental perspective which complicates the valuation of children's HRQOL at different ages and developmental stages, can be included in this approach. So, the necessity to assess the HRQOL of children in the three main domains is acknowledged by most researchers, although there is considerable variability in the choice of subdomains.

Adequate measurement is a paramount issue in the field of childhood HRQOL research.

Several quality of life instruments have been developed during the last ten years. Many instruments measure mainly aspects of physical functioning, especially the functional status of the child. Examples of these instruments are the Play Performance Scale (PPS; Mulhern, Fairclough, Friedman, & Leigh, 1990), the Quality of Well-Being Scale (QWB; Kaplan et al., 1989), and the Functional Disability Inventory (FDI; Walker & Greene, 1991). Some instruments include only a limited number of aspects of psychosocial functioning such as the Multi-Attribute System for Classifying the Health Status of survivors of childhood cancer, in which 'emotion' and 'cognition' are the two psychological attributes next to five attributes concerned with physical functioning and limitations (Feeny et al., 1992). Only a few quality of life instruments cover the psychological and social functioning domains in a more comprehensive way such as the Child Health Questionnaire (CHQ; Landgraf et al., 1998), the Child Quality of Life Questionnaire (CQOL; Graham, Stevenson, & Flynn, 1997), the Munich Quality of Life Questionnaire (KINDL; Bullinger, VonMackensen, & Kirchberger, 1994; Ravens-Sieberer & Bullinger, 1998), and the TNO AZL Children's Quality of Life parent form (TACQOL, Vogels et al., 1998).

Most of these instruments are completed by adults such as parents, caretakers, or clinicians. The need for developing instruments which can be completed by children themselves is stressed by several authors (Eiser, Havermans, Craft, & Kernaham, 1995; Mulhern et al., 1989; Rosenbaum & Saigal, 1996). To date only the KINDL (Bullinger et al., 1994; Ravens-Sieberer & Bullinger, 1998) and the CQOL (Graham et al., 1997) can be completed by children as young as 9 years old.

The goal of the present study was to develop a HRQOL measure which should be applicable to children, which should cover the three broad domains of functioning in children, which could be completed by caretakers as well as the children themselves and which should fulfill the common criteria of reliability and validity. After developing the items for this new HRQOL measure, we tested aspects of its reliability and validity. Reliability requirements for the scales are good internal consistency (Cronbach alpha above .70) and test-retest correlations (Pearson correlation above .70). The convergent validity of an instrument is shown if scales and subscales of that instrument have moderate to high correlations with other measures covering the same aspects of functioning. Because psychological and social functioning are important domains, measures covering several aspects of psychological and social functioning, such as problem behavior, social competence, and cognitive functioning were included in the study. The most important criterion for the validity of a HRQOL instrument is its power to discriminate between groups with different HRQOL. To this end a study group of children with congenital abdominal anomalies was chosen who were expected to have diminished HRQOL compared to children from a healthy control group. In the literature long-term physical problems are reported for children with congenital abdominal anomalies (Chetcuti & Phelan, 1993; Chetcuti, Phelan & Greenwood, 1992; Diseth & Emblem, 1996; Moore, Albertyn, & Cywes, 1996; Vanamo et al., 1996). Concerning their psychological functioning, these children have problems in cognitive functioning (Kato et al., 1993) and they show increased behavioral and emotional problems

(Ginn-Pease et al., 1991; Ludman, Spitz, & Kiely, 1994).

4.3 Methods

4.3.1. Subjects

The sample consisted of two groups of children aged 8 to 12 years. One group, named the study group, consisted of 139 children, 87 boys and 52 girls, who were born in the period from 1982 to 1986 and were treated in a large university children's hospital for a major congenital abdominal anomaly: 31 children with an esophageal atresia, 11 children with a diaphragmatic hernia, 19 children with an abdominal wall defect, 26 children with an intestinal atresia, 23 children with Hirschsprung's disease, 22 children with anorectal malformations, and 7 children with multiple diagnoses. A total of 166 children, which were all the surviving children from the indicated time period, were eligible for this study. Of these, 115 children and the parents of 139 children consented to participate leaving a total response of 83.7 %. The mean age of the study group children at the time of the assessment was 10.6 years (SD 1.1). Most children were hospitalized in the first 3 days of their life. The duration of the first hospitalization ranged from 4 to 690 days (Median 34 days). The children were rehospitalized during their lives between 0 en 29 times (Median 3 hospitalizations) and operated between 0 en 8 times (Median 2 operations). Seventy-seven children (55 %) had only one congenital anomaly, 37 (27 %) had one or more minor associated congenital anomalies such as skeletal or urogenital anomalies, and 25 (18 %) had one or more major associated congenital anomalies such as major cardiac or neurological anomalies.

The second group, named the comparison group, consisted of 136 children from the general population, 83 boys and 53 girls, mean age 10.5 years. The comparison group was selected as follows. The municipal council of six villages and towns with degrees of urbanization corresponding to those of the study group children were approached and they gave names and addresses of a number of randomly selected children with an age and gender distribution corresponding to that of the study group. These children and their parents were approached by telephone to ask their participation and obtain their informed consent. The response rate of the comparison group is estimated to be 60 %. Due to the fact that the municipal councils used different procedures to approach the subjects it is not possible to be more specific about response rate, neither to give information on reasons for refusal. Eventually the comparison group matched with the study group on the variables gender, age, and degree of urbanization.

The distribution of socio-economic status (SES), based on the highest parental occupational level, was as follows for the study group and comparison group respectively: low SES n=25 and n=23; middle SES n=62 and n=54; and high SES n=52 and n=59. In the study group one hundred and seventeen children lived with both parents, 19 children of divorced parents lived with one parent, and 1 child was institutionalized. One hundred and thirty children

in the comparison lived with both parents, and the parents of 5 children were divorced. No children in the comparison group were institutionalized. There were no significant differences between both groups regarding the distribution of either SES or living arrangements.

4.3.2. Measures

4.3.2.1. *The Quality of Life Questionnaire for Children (QLQC)*

Choice of domains, subdomains, and items. In agreement with the majority of childhood HRQOL research we conceptualized HRQOL as a multidimensional construct including the physical, psychological, and social functioning domains (Aaronson, 1991; Landgraf & Abetz, 1996; Mulhern et al., 1989; Spieth & Harris, 1996). The physical functioning domain generally includes disease state or symptomatology and functional status, i.e. the ability to perform age-appropriate daily activities (Spieth & Harris, 1996). There is no clarity in the literature on HRQOL measurement concerning the subdomains which should be included in the psychological and social functioning domains. Inclusion of overt behavioral problems in the psychological functioning domain is recommended by Spieth and Harris (1996), while others mention a positive self-concept or a basic mood of joy (Lindström & Kohler, 1991), or positive and negative emotions (Vogels et al., 1998). In the social functioning domain relations with peers, with family members, and teachers are primarily identified as the most important aspects (Spieth and Harris, 1996).

In agreement with these recommendations, items concerning general physical well-being, physical complaints, and functional status were included in the physical functioning domain of the questionnaire. Items concerning general psychological well-being, emotions, self-concept, and cognitive functioning were included in the psychological functioning domain. Items concerning family functioning, functioning at school, relationship with peers and leisure time activities were included in the social functioning domain. Items were chosen based upon clinical experience, existing childhood questionnaires concerning physical, psychological, and social functioning, and peer-review. Because a HRQOL instrument should be able to measure also positive aspects of functioning (Landgraf & Abetz, 1996; Mulhern et al., 1989), an equal number of items with a positive content (e.g. I am happy) and a negative content (e.g. I am sad) were chosen. Items containing a negation were avoided as much as possible (3 items). The items were phrased simply, so that 8-year-old children would be able to comprehend them.

The first version of the questionnaire, named the Quality of Life Questionnaire for Children (QLQC), contained 108 items with a 3-point scale defined as 0 = seldom or never, 1 = sometimes, and 2 = always or mostly. As time frame, 'the last 2 months' was chosen. The items for the child and parent versions were phrased with the identical words and presented in the identical order.

Pilot-study. Before the actual study was started, a pilot-study was conducted to test if the QLQC could be completed without difficulty by children aged 8 to 12 years. The QLQC was completed by 60 children aged 8 to 12 years from an elementary school. Most of the children were able to complete the questionnaire with minimal supervision in about 15 minutes. Items which were ambiguous or not well understood by the children were removed or rephrased, and items were added to cover the domains in a better way. The a-priori scales showed satisfactory internal consistency. After the pilot-study the QLQC contained 118 items.

4.3.2.2. *Other measures*

The children in the study group and one of their parents were seen in the hospital for a physical examination of the child performed by a pediatric surgeon and a semistructured interview of the parents and children by one of the investigators (ACMvG or NHB) concerning psychosocial functioning of the children. The interview contained questions concerning functioning within the family, at school, and in relation to peers. Problems in these areas were scored as 0 = absent, 1 = light, 2 = moderate, or 3 = severe. A total score was computed by adding these scores. In the same way a total score for the physical examination was computed. This total score reflects problems in physical functioning, i.e. neurological, cardiac, respiratory, abdominal, uro-genital, or orthopedic problems.

Intelligence was tested with two subtests, vocabulary and block design, of the *Wechsler Intelligence Scale for Children-Revised* edition, which together have a correlation of .92 with full scale IQ (Van Haassen et al., 1986).

The *Self-Perception Profile for Children* (SPPC) and the *Abbreviated Depression Questionnaire for Children* (ADQC) were completed by the children. The SPPC (Harter, 1985) is a questionnaire with 36 items with a 4 points scale for 8- to 12-year-old children concerning their self-esteem, and contains six subscales: Scholastic Competence, Social Acceptance, Athletic Competence, Physical Appearance, Behavioral Conduct and General Self-Worth. In a cross-cultural validation study among Dutch children by Van Dongen, Koot, and Verhulst (1993) the factor structure and other psychometric properties presented by Harter were confirmed. The ADQC is a questionnaire with 9 items with a 2 point scale which screens for depression (De Wit, 1987). Based on the total score the child can be rated as 'not depressed' or 'possibly depressed'. The ADQC has good internal consistency and construct validity (De Wit, 1987).

The parents completed the *Child Behavior Checklist* (CBCL; Achenbach, 1991). The CBCL includes a competence and a problem section. The problem section of the CBCL contains 120 items concerning problem behavior which are scored on a 3-point scale. A total problem score can be computed by summing the scores on these items. Two broad-band groupings, Internalizing and Externalizing, and eight syndrome scales were constructed. The competence scales of the CBCL reflect school functioning and social competence of children. A school, social, and activities competence score and a total competence score can be computed. The CBCL has good reliability and validity measures (Achenbach 1991), which were confirmed for

the Dutch translation (Verhulst, Van Der Ende, & Koot, 1996). Based upon CBCL scores children can be divided in children with and children without emotional or behavioral problems. Children with emotional and behavioral problems are defined as children who have scores above the 85.5th percentile on the internalizing and externalizing broad band grouping and total problem score of the CBCL, and children without those problems who have scores below the 85.5th percentile on these CBCL scales. Deviant scores defined in this way provide a clear distinction between children with versus children without psychopathology (Achenbach, 1991; Verhulst et al., 1996). In the same way children with low competence are defined as children with mean CBCL total competence below the 14.5th percentile, and children with normal competence with scores above the 14.5th percentile on this scale respectively.

4.4. Procedure

The research protocol was reviewed and approved by the medical ethical board of the hospital. The children in the study group and comparison group and their parents received a letter in which they were informed about the purpose and procedures of the study. They were contacted by telephone to ask their participation and obtain their informed consent.

The 115 participating children underwent a physical examination, were interviewed and subjected to an abbreviated intelligence test, and they completed the QLQC, SPPC, and ADQC and their parents were interviewed. The participating parents of 139 children completed the QLQC and CBCL at home. Hundred-and-thirty-three children in the comparison group consented to participate and completed the QLQC, SPPC, and ADQC in small groups in a school in the -neighborhood where they lived. The parents of all 136 children in this group completed the same questionnaires as the parents of the study group children at home. Questionnaires were completed by both parents of 120 children in the study group and of 126 children in the comparison group and in all other cases only by the mother. The parents were instructed to complete the questionnaires independently.

At the end of the first assessment procedure 55 children and their parents from both groups consented to complete the QLQC for a second time. The mean interval between the first and second time was two weeks. These parents and children completed the questionnaire at home with the instruction to do this independently. They received separate return envelopes to enhance independent completion.

4.5. Data analysis

The items of the QLQC with a positive content were recoded so that all items had the same sign with a higher score meaning worse functioning.

To construct reliable scales the following steps in the analyses were taken. First, separate exploratory factor-analyses for the physical, psychological, and social functioning domains, using all items within these domains were performed. These exploratory factor analyses were done for each informant (mother, father, and child) apart, using all subjects, study and comparison group together. The criterion to retain a factor after these analyses was that the Eigenvalue should be greater than 1 and that the content of the factor should be interpretable, i.e. should have meaning at face value. For example, if 5 items of a factor with a total of 7 items reflected self-concept this factor was retained, but if in that same factor 2 items regarded self-concept, 3 cognitive functioning, and 2 anxiety this factor was rejected.

The second step was to construct cross-informant factors, i.e. factors with the same items for parents and children. This was done by retaining only those factors which were represented in the factor structure obtained for at least two informants. If an item had a loading of at least .30 on such a factor, this item was included in the cross-informant factor.

The third step was to perform confirmatory factor analyses for every informant apart on this factor structure. For this confirmatory factor analysis the Multitrait Analysis Program (Hays, Hayashi, Carson, & Ware, 1988) was used. Using this program the extent was tested to which each of the items assigned to the cross-informant factors indeed had a loading on this factor for each of the informants but not on other factors. The criterion to delete a certain item from a factor was that in the analyses of 2 or more informants this item would load higher on other factors.

Scales were constructed with the same label and items as the factors which were obtained in this way. Scale scores were computed by adding the scores on the items in the scale, with higher scores indicating worse quality of life. Broad domain scale scores for the physical, psychological, and social functioning domains were computed by adding all the scores of the subscales within one domain. A total score was computed by adding all scale scores.

To assess the reliability of the scales, the internal consistency using Cronbach's alpha, test-retest correlations, and cross-informant correlations, i.e. correlations between mother and father, and between child and mother and father respectively, were computed. Paired samples t-tests were performed to analyze differences between scores of different informants.

The discriminant validity of the scales was assessed by analyzing the differences between the study group and the comparison group using two sample t-tests. Those children of the comparison group with a chronic disease or a congenital anomaly ($n = 12$) were excluded from these analyses. Further, two-sample t-tests were performed to analyze the differences in QLQC scale scores of parents and children between children with and without emotional and behavioral problems, between children with low and normal social competence, and between children with and without possible depression. For these analyses mean scores on the CBCL and QLQC scales for the parents were computed by adding the scores of the mother and father and dividing those by two. If only the mother had completed the questionnaires her scores were given.

The convergent and divergent validity of the QLQC was tested by computing Pearson-correlations between scores on the QLQC and scores based on the physical examination,

psychosocial interviews, intelligence, mean scores on the CBCL, and scores on the SPPC which reflect similar constructs as those tapped by the QLQC.

4.6. Results

4.6.1. Scale construction

The exploratory factor analyses yielded an identical factor structure for each of the three informants with three factors in the physical functioning domain, four factors in the psychological functioning domain, and six factors in the social functioning domain for the children and five factors for the parents, because a consistent factor named 'Relations with parents' appeared only in analyses of the child data. In the confirmatory factor analysis more than 90 % of the items had factor loadings higher than .30 for all informants. Two items had to be deleted from factors because they had higher cross-loadings on several other factors with at least two informants. Two items were deleted from that factor to which they were assigned and included in another factor because they had a higher loading on that last factor with all three informants. One item was deleted from that factor to which it was assigned because with two informants the corrected item-total correlation was below .30. These factors yielded the 13 scales of the QLQC. In table 4.1 the scales with the number of items and examples of items are given.

Table 4.1

QLQC-scales with number of items in the scales and examples of items

QLQC scales	n	Examples of items
<i>Physical functioning</i>	18	
Physical complaints	9	I have stomach ache. I am dizzy.
Physical limitations	6	I am good at sports or gym. I cannot participate in sports because of physical problems.
Physical handicaps	3	I can walk well. I can see well.
<i>Psychological functioning</i>	32	
General well-being	7	I am satisfied with my life. I have pleasure in my life.
Cognitive functioning	10	I am good at learning. I can keep my attention with my schoolwork.
Self-concept	7	I find that I do most things well. I have self-confidence.
Anxious-depressed feelings	8	I am feeling sad. I am afraid.
<i>Social functioning</i>	36	
Relation with parents (children only)	4	I have confidence in my parents. I am feeling well in my family.
Relation with siblings	3	I can play/talk well with my brother(s)/sister(s). I am helped by my brother(s)/sister(s) if necessary.
Relation with peers	8	I have enough good friends. Other children like to play with me.
School functioning	9	I like to go to school. I behave well at school.
Social conflicts	8	I quarrel with my parents. I tease other children.
Leisure-time activities	3	I am bored. I can keep myself busy during leisure time

4.6.2. Reliability of the scales

4.6.2.1. Internal consistency.

Internal consistency of the scales as indicated by Cronbach's alpha is given in table 4.2 for mother, father, and child. Almost all alpha's of the broad domain scales were above .80 (.76 to .92, Median .87). Seventy-five percent of the alpha's of the subscales were above .70 (.60 to .90, Median .75). The weakest scales for all informants were those within the social functioning domain, especially Relations with Parents and Siblings (Cronbach's alpha .60 to .68), and School Functioning (Cronbach's alpha .63 to .69). Dispersed across different subscales, 5 items for the child, 3 items for the father, and 1 item for the mother had a corrected item-total correlation with

its own scale between .19 and .30. Five of these items, mainly in the psychological functioning domain scales, had corrected item-total correlations higher than .40 with the two other informants, and the other four, mainly in the social functioning domain scales, had corrected item-total correlations between .30 and .43 with the other two informants. All other items had corrected item-total correlations higher than .30 with all three informants.

Table 4.2

Cronbach's alphas and test-retest correlations of the QLQC scales, for mothers, fathers, and children

QLQC scales	mother		father		child	
	α (268)†	r_{tt}^* (55)	α (246)	r_{tt} (52)	α (243)	r_{tt} (54)
<i>Physical Functioning</i>	.77	.59‡	.83	.54	.76	.55
Physical Complaints	.72	.41§	.85	.36	.75	.55
Physical Limitations	.80	.43	.80	.80	.77	.42
Physical Handicaps	.82	.43	.85	.41	.62	.61
<i>Psychological functioning</i>	.92	.94	.91	.79	.88	.85
General Well-Being	.90	.80	.88	.78	.78	.68
Cognitive Functioning	.86	.92	.85	.84	.73	.82
Self-Concept	.86	.85	.82	.66	.70	.79
Anxious-Depressed feelings	.75	.89	.73	.74	.74	.72
<i>Social functioning</i>	.87	.89	.86	.83	.87	.74
Relation-Parents	-	-	-	-	.68	.61
Relation-Siblings	.67	.77	.60	.57	.64	.60
Relation-Peers	.90	.88	.85	.88	.80	.70
School Functioning	.69	.70	.66	.71	.63	.67
Social Conflicts	.78	.74	.79	.68	.74	.78
Leisure-Time Activities	.71	.59	.72	.68	.64	.54

* r_{tt} = 2-week test-retest Pearson correlations. † Number of subjects in analysis.

‡ All test-retest correlations were significant at $p < .001$, except § $p < .01$.

4.6.2.2. Test-retest correlations.

As is shown in table 4.2 the test-retest correlations within the physical functioning domain were lower than .70 with one exception (.36 to .80, Median .48). Ninety percent of the test-retest correlations within the psychological functioning domain were higher than .70 for all informants (.54 to .94, Median .80). In the social functioning domain all test-retest correlations for the broad domain scale, the Peer Relations, and the Social Conflicts scale except one were higher than .70 (.68 to .89, Median .78), but for the other subscales in this domain 70 % of the test-retest correlations were lower than .70 (.54 to .77, Median .67). In general the highest test-retest correlations were found for the mothers (.41 to .94, median .77) and the lowest for the children (.42 to .85, Median .67).

4.6.2.3. Correlations between informants.

Correlations of scale scores between each pair of informants are shown in table 4.3. The correlations between informants were low to moderate in the physical functioning domain (.04 to .75, Median .32) and moderate to high in the psychological and social functioning domains (.24 to .85, Median .45). All correlations between the parents (.13 to .85, Median = .72) were higher than those between the parents and the child (.04 to .51, Median = .37). The scores of the children were higher than those of the parents on two-thirds of the scales with effect-sizes, expressed as the percentage of variance explained by the group effect, between 2 % to 15 %. The mothers scored higher than the fathers on most scales of the psychological functioning domain and on the Relation with Siblings scale, although the effect-sizes of these differences did not exceed 5 % explained variance

Table 4.3

Pearson correlations and effect-sizes of paired t-tests between scores of different informants

QLQC scales	Mother-Father (n = 244)		Mother-Child (n = 240)		Father-Child (n = 220)	
	r*	η^2 †	r	η^2	r	η^2
<i>Physical functioning</i>	.56	-	.37	.09	.38	.07
Physical complaints	.41	-	.25	.10	.29	.11
Physical limitations	.75	-	.36	-	.29	-
Physical handicaps	.13‡	-	.22	-	.04§	-
<i>Psychological functioning</i>	.85	.04	.51	-	.44	.03
General well-being	.73	.02	.41	.03	.29	.08
Cognitive functioning	.85	-	.51	-	.49	-
Self-concept	.75	.05	.34	.02	.27	-
Anxious-depressed	.67	.02	.38	.02	.36	-
<i>Social functioning</i>	.79	.02	.47	.08	.38	.13
Relation-parents	-	-	-	-	-	-
Relation-siblings	.61	.03	.35	.07	.28	.12
Relation-peers	.77	-	.49	-	.42	-
School functioning	.72	-	.41	.15	.34	.02
Social conflicts	.66	-	.35	-	.33	-
Leisure-time activities	.60	-	.30	.03	.24	.15

* r = Pearson correlations; all correlations significant at $p < .001$ except ‡ $p = .04$ and § $p = ns$.

† η^2 = proportion of variance explained by differences between informants in paired t-tests.

4.6.3. Influence of demographic and medical background variables on quality of life

Boys obtained higher scores than girls on several scales in the psychological and social functioning domain as reported by the children and by the parents ($p < .001$ to .023). The effect-sizes were in most cases lower than 4 % explained variance. There were few differences between

boys and girls within the physical functioning domain. Based on the children's self-reports, the children with low SES obtained on half of the scales of the QLQC higher scores than children with high SES, also with effect-sizes below 4 % explained variance ($p < .01$ to $.03$). This difference between low and high SES was not found in the parent reports.

4.6.4. Validity of the scales

4.6.4.1. Discriminative validity

Table 4.4 shows mean scores and standard deviations on those scales that showed significant differences between the mean scores of the study group versus the comparison group. In the physical functioning domain, children and mothers in the study group indicated worse overall functioning. Mothers and fathers reported more physical limitations for their children while the children themselves reported more physical complaints. In the psychological functioning domain all informants reported worse cognitive functioning of the children in the study group.

Children in the study group reported more anxious-depressed feelings, and in the social functioning domain more problems in functioning at school than children in the comparison group. The children in the comparison group reported more problems in their relations with siblings. The effect-sizes must be regarded as rather small, because the effect-sizes did not exceed 5 % explained variance for most effects (cf. Cohen, 1988).

Within the study group there were no differences in QLQC scores between the children in the different diagnostic groups. However, children with major associated congenital anomalies obtained higher scores on the broad physical functioning domain scale (Mean 6.62) than children without (Mean 3.71, $p < .01$) as reported by the parents. Number of hospitalizations and number of operations correlated moderately ($r .25$ to $.35$, all p -values $< .01$) with the score on the broad physical functioning domain scale as reported by the parents. No relations between these medical background variables and the scores on scales in the psychological, and social functioning domains were found.

The mean scores on the broad psychological and social functioning domain scales and the total score on the QLQC of the parents of children with behavioral/emotional problems were two to 2.5 times higher than those of children without behavioral/emotional problems (Table 4.5). The QLQC scores of the children themselves in the same domains were about 1.5 times higher for children with high levels of problem behavior versus children without (Table 4.5). All comparisons except one were significant, also after Bonferroni correction for multiple comparisons ($p < .001$). The scores on the broad social functioning domain scale of the QLQC were 1.5 to 2 times as high for children with low total competence versus those with normal total competence on the CBCL (Mean low competence group = 15.1 to 21.7 versus Mean normal competence group = 9.0 to 11.9, all p -values $< .001$).

Table 4.4

Significant differences between study group and comparison group on QLQC scales

QLQC scales	Study group		Comparison group		p	η^2 *
	M	SD	M	SD		
<i>Mother</i>	(n = 132)		(n = 125)			
Physical functioning	4.25†	3.92	2.86	3.17	.002	.04
Physical limitations	1.30	2.10	0.48	1.22	.000	.05
Cognitive functioning	4.67	4.06	3.48	2.94	.008	.03
<i>Father</i>	(n = 120)		(n = 115)			
Physical limitations	1.25	1.93	.77	1.77	.046	.02
Psychological functioning	10.75	8.13	8.47	7.13	.024	.02
Cognitive functioning	4.64	3.79	3.09	2.97	.001	.05
<i>Child</i>	(n = 110)		(n = 121)			
Physical functioning	5.42	4.23	4.09	3.33	.009	.03
Physical complaints	3.77	3.09	2.86	2.15	.010	.03
Psychological functioning	12.45	7.7	10.44	7.24	.043	.02
Cognitive functioning	4.48	3.08	3.59	2.77	.022	.02
Anxious-depressed feelings	4.47	2.84	3.61	2.53	.017	.02
Relation-siblings	1.63	1.34	2.05	1.37	.028	.02
School functioning	2.76	2.15	2.08	2.11	.016	.02

* η^2 = proportion of variance explained by differences between groups.

† Higher scores indicate worse functioning.

The children's and parents' scores on the QLQC Anxious-Depressed scale for children with possible depression were twice as high as those for children without possible depression (Children: with possible depression Mean = 7.9 [n = 22], without possible depression Mean = 3.6 [n = 220]; $p < .001$; Parents: with possible depression Mean = 6.2 [n = 24], without possible depression Mean = 3.2 [n = 218], $p < .001$).

4.6.4.2. Convergent and divergent validity

Moderate to high correlations were found between scores obtained from the physical examinations of the children and the interviews with the children and parents on the one hand and relevant QLQC scores on the other hand. In the physical functioning domain, the total score of the physical examination correlated moderately with the mean broad physical functioning domain scale score of the parents ($r = .50$), but low with this score of the children ($r = .21$). The total score of the interviews concerning psychosocial functioning correlated highly with the broad psychological and social functioning domain scores for the children as well as the parents ($r = .62$ to $.65$). Further, moderate to high correlations between scores on scales of the QLQC and comparable scales of the CBCL, and SPPC, and IQ were found (Tables 4.6 and 4.7). For example, the correlation between the Physical Complaints scale of the QLQC and the Somatic Complaints syndrome of the CBCL was $.56$, and $.71$ between the anxious-depressed scales of

both QLQC and CBCL.

Table 4.5

Mean scores on the QLQC broad psychological and social functioning domain scales and total score of children with mean parental CBCL total problem scores in the normal versus the borderline range.

QLQC scales	CBCL Total Problem score	
	Normal	Borderline
	M (SD)	M (SD)
<i>Parent reports</i>		
Psychological functioning	7.7 (5.4)	19.9 (9.2)
Social functioning	7.4 (4.0)	16.1 (6.9)
Total score	18.1 (9.5)	41.6 (16.4)
<i>Child reports</i>		
Psychological functioning	10.3 (7.0)	15.4 (8.5)
Social functioning	10.7 (6.7)	15.7 (7.2)
Total score	25.1 (14.5)	37.6 (16.8)

Note. Independent samples t-tests between normal and borderline groups were significant at $p < .001$. The number of children in the normal groups varied from 162 to 228 and in the borderline groups from 30 to 69.

The correlation between the Self-Concept scale of the QLQC and the General Self-Worth scale of the SPPC was $-.62$, and $-.65$ between the Cognitive Functioning scale of the QLQC and the School-Competence scale of the SPPC. Correlations between scales which were supposed to assess unrelated aspects of functioning were low to moderate. Confirmation for the convergent validity of the Cognitive Functioning scale of the QLQC was obtained from the moderate negative correlation between that scale and mean WISC-R IQ ($r = -.32$, $p < .001$) while all other correlations between IQ and QLQC scales were not significant.

Table 4.6

Pearson correlations between the mean scores of mothers and fathers on relevant QLQC and CBCL scales and IQ.

QLQC scales	CBCL syndromes					IQ
	Somatic Complaints	Anxious-Depressed	Aggressive Behavior	Social Problems	Attention Problems	
Physical complaints	.56*†	.31	.16§	.26	.24	-.17¶
Cognitive functioning	.19†	.35	.39	.55	.71	-.38
Anxious-depressed	.41	.71	.43	.55	.53	-.04¶
Peer-relations	.11¶	.58	.49	.68	.51	-.01¶
Social conflicts	.14¶	.48	.68	.49	.42	-.07¶

Note. Only QLQC and CBCL scales are included that were expected to show high cross-correlations.

* Fat printed correlations concern comparable scales.

† All p-values are $< .001$, except ‡ $p < .01$, § $p < .05$, ¶ $p = ns$.

Table 4.7

Pearson correlations between relevant QLQC and SPPC scales.

QLQC scales	SPPC-scales			
	School Competence	Social Acceptance	Behavioral Conduct	General self-worth
General Well-Being	-.28*	-.49	-.38	-.58†
Cognitive Functioning	-.65	-.36	-.55	-.47
Self-concept	-.37	-.43	-.42	-.62
Peer-relations	-.30	-.73	-.39	-.43
Social Conflicts	-.23	-.28	-.46	-.31

Note. Only QLQC and SPPC scales are included that were expected to show high cross-correlations

* All p-values are < .001. † Fat printed correlations concern comparable scales.

4.7. Discussion

The QLQC is a 118 item self-report and parent-report generic health-related quality of life questionnaire for children covering the broad domains of physical, psychological, and social functioning. Within these domains three to six subscales could be discriminated. Identical scales for parents and children were constructed. The present study showed that the psychometric properties of the instrument are promising as indicated by adequate reliability and validity measures for several scales, but that further study with the instrument is needed.

The QLQC has been tested with a group of 139 children aged 8 to 12 years with major congenital abdominal anomalies and a matched comparison group of 136 children. The children in the study group were characterized by a large number of physical problems early in life such as a long hospitalization in the newborn period, many operations, and a large proportion of children with associated congenital anomalies.

The QLQC could easily be administered to children of this age group. Almost all children understood the questions well, and in spite of the rather large number of items most children enjoyed completing the questionnaire. The completion of the QLQC took the children about 15 minutes. The QLQC has been tested in this study on a population of 8- to 12-year-old children, but it also might be applicable for younger children, from 6 years on, as parent-report questionnaire, and for adolescents as self-report as well as parent-report questionnaire.

The reliability of the broad domain scales and several subscales in the psychological and social functioning domain was good as is shown by good internal consistency and test-retest correlations. In the physical functioning domain the test-retest correlations were rather weak. This may be caused by the rather small variance of the physical functioning scales so that small changes in the item scores over time may have had a relatively large effect on the scale scores. This means that the physical functioning scales of the QLQC need further improvement to enhance the reliability of the scales and testing of their reliability in a sample that yields more variance in scores. The scales with the weakest internal consistency were the Parents and Siblings

Relations scales and the School Functioning scale. The number of items in the first two scales may have been too small ($n = 3$ and $n = 4$) to yield reliable scales for measuring relations within the family. An explanation for the low internal consistency of the School Functioning scale may be that it contained rather diverse items such as behavior at school, relationship with teacher, and feelings about school. Also, the corrected item-total correlations of 4 items were lower than .30 with one informant while the item-total correlation with the other two informants were not higher than .43. This is an indication that these items did not add sufficiently to the internal consistency of the concerning scales. This means that in a later version of the QLQC these items might be deleted and that items should be added to several scales to enhance reliability.

The correlations between the scores of the parents in the psychological and social functioning domains were between .60 and .85, which is within the expected range (Achenbach, McConaughy & Howell, 1987). The correlations in all domains between the scores of the children and the parents were much lower (.04 to .51) and the scores of the children were consistently higher than the scores of the parents. In several other HRQOL studies large differences between the ratings of parents and children were found (Guyatt, Juniper, Griffith, Feeny, & Ferrie, 1997; Theunissen et al., 1998). In a recent study concerning this 'proxy-problem' Theunissen et al. (1998) showed that the children's views cannot be put aside as being invalid due to their supposed inability to evaluate their HRQOL. Using multitrait-multimethod analyses they showed that the ratings of children aged 8 to 11 years old were as valid as those of the parents. Thus, the hypothesis is confirmed that children have a different but valid view on their own functioning than their parents. This underlines the necessity of the present effort to develop a self-report health-related quality of life instrument for children.

An important aspect of a quality of life instrument is its power to discriminate between groups with different levels of quality of life. We expected that children with congenital abdominal anomalies would have a lower level of quality of life than children in the comparison group. This hypothesis was partly confirmed. We found that children in the study group had more problems in physical functioning than children in the comparison group as reported by the children and the parents, although the effect sizes for these differences were small. Within the study group, children with major associated anomalies, more operations, and hospitalizations obtained higher scores in the physical functioning domain as reported by parents. Because this group with more problematic medical background was expected to have more problems in long-term physical functioning, which was confirmed during the medical examinations, this result can be seen as a confirmation of the discriminative validity of the QLQC. In the psychological functioning domain, the scores on the Cognitive Functioning scale of the parents as well as the children in the study group were lower than those in the comparison group. On other aspects of functioning the QLQC detected only a few differences between the study group and the comparison group. On the QLQC, children in the study group reported more anxious-depressed feelings and more problems at school. Apart from cognitive functioning, the parents did not report worse psychological or social functioning on the QLQC for the children in the study group.

There may be several explanations for the few differences found between the study group and the comparison group and between the different diagnostic groups.

1. *The discriminative validity of the QLQC is insufficient with the effect that real differences in HRQOL were not detected by the QLQC.*

This explanation can be questioned because with other assessment methods such as the physical examinations and the well validated standardized questionnaires on the children's functioning few differences were found between the study group and comparison group children (Bouman et al., in preparation). Further, to several real effects the QLQC proved to be sensitive (i.e.: the difference in IQ between the study group children and the norm, the difference in psycho-social functioning between children with and without emotional and behavioral problems, between children with and without low social competence, and between children with and without possible depression).

2. *The HRQOL of children with congenital abdominal disorders is not very much reduced in comparison to healthy children.*

This is confirmed by the fact that differences in mean scores on the CBCL, SPPC, and ADQC between the study group and the comparison group were rather small or absent.

3. *There are few real differences between children with different congenital abdominal anomalies.*

Again, other instruments such as the CBCL, SPPC, and ADQC neither discriminated between diagnostic groups in this study (Bouman et al., in preparation). There are other studies in which there were found only few differences between children with different congenital abdominal anomalies. For example, Ginn-Pease et al. (1991) failed to find a hypothesized difference in psychosocial functioning between children with abdominal wall defects and children with anorectal malformations. Lastly, there are strong indications that general aspects of diseases have more influence on HRQOL than disease specific aspects (Wallander & Varni, 1998). Therefore it can be doubted that differences between diagnostic groups are a priori to be expected.

4. *As a generic HRQOL instrument the QLQC is not sensitive enough for the specific problems of children with congenital abdominal anomalies.*

There are indeed some specific physical problems such as swallowing problems for children with esophageal atresia or problems with continence for children with anorectal malformations which are not detected by the QLQC. However, these problems were not related to worse psychosocial functioning in our study group (Bouman, Koot, & Hazebroek, in press; Bouman, Koot, Langemeijer, & Hazebroek, in press).

It may be concluded that there are indications of good discriminative validity of the QLQC in several aspects, but that further confirmation of the discriminative validity the QLQC is needed. To this end in a future study with the QLQC it will be needed to incorporate groups with more reduced HRQOL than the children in this study group and to compare their HRQOL with that of healthy controls.

The correlations between scales of the QLQC with scores from the physical examinations of the children and the psychosocial interviews, and comparable scales of other well-validated instruments were all moderate to high while the correlations between non-converging scales of the QLQC and CBCL were low to moderate. This means that there is an overlap of 30 % to 50 % shared variance between QLQC measures and other well validated measures for several aspects of functioning which is an indication of good convergent validity of those QLQC scales.

Mulhern et al. (1989) recommended that a HRQOL index should (a.) measure physical as well as emotional and social aspects of a child's functioning; (b.) be sensitive to functional problems; (c.) be reliable and valid; (d.) be brief, simple and easy to administer; (e.) depend on ratings of caretakers; (f.) have age-corrected general population norms; (g.) allow for supranormal performance; (h.) permit reliable estimates for premorbid functioning; and (i.) permit children to give self ratings. The QLQC partly fulfills at least seven of these criteria (a, b, c, d, e, g, and i). The QLQC assesses the subjective appraisal of a child's functioning by child and parents and it does this in a homogeneous way across informants so that information from parents and children can adequately be compared. It is a short measure which can easily and repeatedly be completed by children and parents during hospital visits . If norms are obtained, these have the advantage of providing norms obtained in one normative group for different aspects of functioning, the assessment of which would otherwise require different instruments each having their own norm groups (e.g. CBCL, SPPC etc.). Although the QLQC has several good psychometric properties, further research with the QLQC is needed. Several adaptations will be necessary to enhance its reliability and validity. Future research with a renewed version of the QLQC on other patient populations with more actual problems in physical, psychological, and social functioning will reveal its capacity to fulfill the existing need of generic HRQOL self-reports for children.

Chapter 5

Children with congenital diaphragmatic hernia are at risk for lower levels of cognitive functioning and increased emotional and behavioral problems

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5.1. Summary

The physical, psychological, and social functioning of 11 children aged 8 to 12 years with Congenital Diaphragmatic Hernia (CDH) was assessed with several standardized assessment procedures. Physically most children functioned well at follow-up with half of the children showing minor physical problems such as bronchial hyperreactivity. The mean IQ of the children was 15 points (1 SD) below the norm of 100. Only 6 children were at expected school level. The children showed more emotional and behavioral problems than in the general population as reported by parents and teachers. The children themselves reported more depressive problems, but not a lower self-esteem than children in the general population. These results were confirmed by the results of interviews with parents and children concerning psychosocial functioning.

It is concluded that children with CDH show more cognitive and learning problems and increased rates of emotional and behavioral problems compared to children in the general population. Since no children treated with Extra Corporeal Membrane Oxygenation (ECMO) were involved in this study, the earlier reports that lower cognitive functioning is limited to children with CDH treated with ECMO can not be confirmed by this study. Considering the results of this study there is a need for further follow-up studies concerning the long-term psychological and social functioning of children with CDH.

5.2. Introduction

Congenital Diaphragmatic Hernia (CDH) is a life-threatening congenital anomaly which, notwithstanding developments in treatment modalities, has an overall mortality rate of 40 to 60 percent (Langham et al., 1996). Until the late eighties children with CDH were operated immediately after admission, but now surgery is delayed several days until the clinical condition of the infant is stabilized (Davenport, Rivlin, D'Souza, & Bianchi, 1992). Treatment after surgery may be prolonged and characterized by many complications. In recent years more attention has been paid to the long-term follow-up of these children. Although underlying structural pulmonary problems, such as remaining hypoplasia, gastroesophageal reflux, poor growth, chest-wall asymmetry, and scoliosis are possible long-term complications, many survivors of CDH have good health and no functional limitations (Nobuhara, Lund, Mitchell, Kharasch, & Wilson, 1996; Van Meurs et al., 1993). Very little attention has been paid to the psychological and social functioning of patients with CDH. Since the use of Extra Corporeal Membrane Oxygenation (ECMO) for patients with CDH several studies reported developmental delays in these children (D'Agostinho et al., 1995; Lund et al., 1994, Stolar, Crisafi, & Driscoll, 1995). There are no follow-up studies of children with CDH focusing on other aspects of psychological and social functioning, although problems in these areas can be expected because of the seriousness of the initial physical condition, the long and intensive treatment in the newborn period and the long-term physical sequelae. In this study, we assessed the physical, psychological, and social functioning of 11 children aged 8 to 12 years old with CDH.

5.3. Materials and methods

5.3.1. Participants

The study population consisted of 11 children with CDH, 6 boys and 5 girls, aged 8.4 to 11.8 years (mean age 10.4 years). The children were all hospitalized in the pediatric surgical intensive care unit on the first or second day of their lives. According to the standard procedure at that time, ten children were operated directly while for one child surgery was delayed several days. The length of the first stay in hospital ranged from 10 to 180 days (median 53.7 days). The duration of artificial ventilation period ranged from 1 to 143 days (median 10 days). Three children developed bronchopulmonary dysplasia, defined as the need for supplemental oxygen after the 28th day of life. Five children who were hospitalized within one day after birth because of progressive respiratory insufficiency and were ventilated more than 10 days (12 to 143 days), were considered as high-risk children and the others as low-risk children. Two children were hospitalized only once and nine children were rehospitalized between 1 and 10 times. Eight children were operated once and 3 children underwent more than one laparotomy related to the

CDH. None of the children were treated with ECMO. Five children had no associated congenital anomalies. Four children had minor associated anomaly such as an open ductus Botalli, and two children had a major associated anomaly such as a ventricular septum defect.

Nine children lived with both parents. The parents of two children were divorced and these children were living with their mothers. The socio-economic status (SES) of the families, based on the highest occupational level in the family (Van Westerlaak, Kropman, & Collaris, 1975), was as follows: low SES: n=2; middle SES: n=6; high SES: n=3.

5.3.2. Methods

The children and one of their parents were seen in the hospital for follow-up examinations. A pediatric surgeon performed a general physical examination of the children. In addition, the results of a recent follow-up study concerning pulmonal functioning by IJsselstijn, Tibboel, Hop, Molenaar, and De Jongste (1997), in which a medical history, physical examination, and lung-function tests were performed, were available for all patients. A semi-structured interview on functioning within the family, at school and in relation to peers was taken from the child and one of the parents by a psychologist or child psychiatrist. Problems in these domains were scored on a 4-point scale with 1 = absent, 2 = light, 3 = moderate, and 4 = serious.

IQ of the children was tested with 2 subtests, Vocabulary and Block Design, of the Wechsler Intelligence Scale for Children (WISC-RN) which together have a correlation of .92 with full scale IQ (Van Haassen et al., 1986). Adaptive behavior in three domains, 'communication', 'daily living', and 'socialization' was assessed with the Vineland Adaptive Behavior Scales (VABS) (Sparrow, Balla, & Cicchetti, 1984). Scale scores for each of these domains and a total score can be computed. The VABS have good psychometric properties (Sparrow et al., 1984). No Dutch norms were available at the time of the investigation.

Children completed the Self-Perception Profile for Children (SPPC) a 36-item questionnaire for 8-12 year old children measuring self-esteem. The reliability and validity of the original 6 scales, General Self-Worth, Physical Appearance, Athletic Competence, Scholastic Competence, Social Acceptance, and Behavioral Conduct, were confirmed for the Dutch translation (Van Dongen, Koot, & Verhulst, 1993). The children also completed the Abbreviated Depression Questionnaire for Children (ADQC) a 9-item questionnaire screening for depression which has good internal consistency and construct validity (De wit, 1987).

Parents completed the Child Behavior Checklist (CBCL) to assess behavioral and emotional problems of the children (Verhulst, Van Der Ende, & Koot, 1996). The CBCL contains 120 items which are scored on a 3-point scale with 0 = not true, 1 = somewhat or sometimes true, and 2 = very true or often true. A total problem score is computed by summing all 0s, 1s, and 2s. Two broad band groupings are scored, the Internalizing grouping including withdrawn behavior, somatic complaints without physical cause, and anxious-depressed feelings, and the Externalizing grouping including aggressive and delinquent behavior. The good reliability and validity of the

CBCL were confirmed for the Dutch translation (Verhulst et al., 1996). The teachers of the children completed the Teacher's Report Form (TRF) which is a teacher version of the CBCL. The problem behaviors are scored on the same broad band groupings as in the CBCL. As with the CBCL, the reliability and validity of the TRF have been confirmed for the Dutch version (Verhulst, Van Der Ende, & Koot, 1997).

5.4. Procedure

The study was reviewed and approved by the medical ethical review board of the hospital. The children and their parents received a letter in which they were informed on the purpose and procedures of the study. They were then contacted by telephone to ask their participation and to obtain their informed consent. Of the 16 eligible children, 11 participated (69 %). The fact that most children had recently participated in a follow-up study of pulmonary function probably influenced the response negatively. Ten children and their parents visited the hospital for interviews and completion of the questionnaires by the children. The parents of one child completed the questionnaires, but were not interviewed. The mother, and also the father if present, completed the questionnaires at home. The parents were instructed to complete the questionnaires independently. The teachers of the children were sent a questionnaire which was returned in all cases. The results of the questionnaires will be presented for all 11 children, and the results of the interviews and IQ tests for the 10 children, who were interviewed and tested.

5.5. Statistical procedures

The results of the interviews are given as frequencies. Range and means of IQ and VABS scores are given. Further, based on Dutch and American norms respectively, IQ and VABS scores are divided in scores in the normal range, i.e. scores of 85 or higher, reflecting normal cognitive or adaptive functioning, or in the borderline range, i.e. scores below 85, reflecting borderline cognitive or adaptive functioning. For the CBCL and TRF problem scores frequencies of children with scores in the normal and borderline range are given. Based on Dutch normative data, the borderline range is defined as problem scores above the 85.5th percentile for the Total Problems, Internalizing, and Externalizing scores. Deviant scores defined in this way provide the strongest possible distinction between children with psychopathology versus children without (Verhulst et al., 1996). Non-parametric binomial tests were performed to assess if the distribution of the frequencies differed from expectations. Mean scores on the SPPC were compared with mean scores in the general population (Van Dongen et al., 1993), and frequency of possible depression was computed based upon general population norms of the ADQC (De wit, 1987). All significance levels were fixed at $p < .05$.

5.6. Results

The physical examination showed that most children functioned well physically. Five children were completely healthy. Three children reported complaints of bronchial hyperreactivity and had some reduction in exercise tolerance during a recent study one year before the present follow-up (IJsselstijn et al., 1997). Two of them still had some complaints. Five children had signs of gastroesophageal reflux when they were younger, but none had complaints presently. One of these children had been operated with a Nissen fundoplication at the age of 10. Five children had slight miscellaneous complaints such as headache, abdominal pains, and pains in arm or legs. One child was impaired to participate in sports by a congenital hip dislocation. No children had serious physical impairments or handicaps related to CDH.

The mean IQ of the children was 85. This is 15 points or one standard deviation lower than the norm of 100 (Table 1). Significantly more children than expected obtained IQ scores in the borderline range. Six of the children were at the expected school-level for their age, two children were one class behind, and three children required special education. The results of the adaptive behavior scales were comparable with the IQ scores of the children. The mean total VABS score was 87, which is 13 points or .9 SD lower than the norm of 100 (Table 5.1). One boy was classified as mentally retarded because he had an IQ score and total VABS score below 70. There were no differences between the high-risk and low-risk children in intelligence, school level, or adaptive functioning.

Table 5.1

Mean IQ-scores and VABS* domain scores and total score, and numbers of children (n = 10) with scores in the normal versus the borderline range for IQ and VABS

	Mean	range	≥85	< 85	p†
IQ	85	54 - 106	5	5	.009
VABS Domains:					
Communication	87	66 - 120	6	4	.05
Daily living	91	54 - 114	8	2	ns
Socialisation	89	48 - 115	8	2	ns
Total score	87	54 - 110	8	2	ns

* Vineland Adaptive Behavior Scales

† Binomial tests of observed versus expected proportion of scores 1 SD below the norm.

The children showed significantly more emotional and behavioral problems than expected as reported by the parents and the teachers on the CBCL. Five children according to the mothers and 6 children according to the teachers had scores in the borderline range on the CBCL and TRF total problemscore which is significantly more than expected. According to the mothers significantly more children than expected obtained scores in the borderline range on the

Internalizing broad band grouping ($n = 6$), while according to the fathers significantly more children obtained scores in the borderline range on the Externalizing broad band grouping ($n = 4$).

Seven children were rated as 'not depressed', three children as 'possibly depressed', and one child as 'probably depressed'. These results indicate that more children than expected had depressive problems according to their own reports. The mean scores of the children on the scales of the SPPC were within the normal range indicating that most children did not have low self-esteem. One child had a very low self-esteem as indicated by scores below the normal range on five of the six scales of the SPPC. This child was also rated as probably depressed. Concerning these indicators of psychosocial functioning there were no differences between the high-risk and low-risk children.

The results of the interviews concerning psychosocial functioning indicated that six children did well. They had no or only slight problems in the family, at school, or in the interaction with peers, according to parents as well as themselves. Parents indicated that two children had moderate problems. One child was not functioning well at school and probably would be placed at a lower school level. One child had some problems in the interaction with her siblings and showed some psychosomatic complaints such as migraine and constipation. Two children had serious problems as indicated by the parents as well as themselves. They had problems at home, at school, and with peers. Both children had received help from a mental health service in the past. The mother of one of these children considered placement of her child in an institution. At the time of the investigation no children received help from a mental health service.

5.7. Discussion

Although there is a growing interest in the long-term physical as well as psychological and social functioning of children with congenital abdominal anomalies (Kato et al., 1993; Ludman & Spitz, 1995; Tarnowski, King, Green, & Ginn-Pease, 1991), this is the first study in which all these aspects of functioning were assessed for children with CDH. No children had serious health problems or impairments. These results are comparable to the results of other follow-up studies (Nobuhara et al., 1996; Stolar, 1996).

The children in the present study showed a reduced level of cognitive functioning and school functioning. Only five children had IQ scores in the normal range and only six children were at expected school level. No differences concerning these parameters were found between high-risk children and low-risk children. A number of studies have shown that especially children with CDH treated with ECMO are at risk for cognitive developmental delays (D'Agostino et al., 1995; Lund et al., 1994; Stolar et al., 1995; Van Meurs et al., 1993). Only two studies reported on the cognitive development of CDH children not treated with ECMO (Davenport et al., 1992,;

Nobuhara et al.,1996). Davenport et al. (1992) reported that none of 23 children with CDH aged 18 to 94 months, not treated with ECMO but with delayed surgery, had developmental delays on the Griffiths's mental developmental scales. Based on the results of non-specified developmental examinations Nobuhara et al.(1996) concluded that children with CDH treated with ECMO more frequently had developmental delays than CDH children not treated with ECMO. Combination of the results of these studies suggests that only children with CDH who need ECMO are at risk for cognitive problems. However, this conclusion does not seem justified because in our study none of the children were treated with ECMO, while the children showed clearly a reduced level of intellectual and school functioning. We conclude that the whole group of children with CDH may be at risk for cognitive delays. These cognitive delays are possibly related to several factors which may negatively influence central nervous system development such as hypoxia, hypercapnia, and acidosis which are frequent problems in children with CDH either in the immediate postnatal period or during the course of stay in the intensive care unit.

We found that children with CDH showed elevated levels of emotional and behavioral problems as reported by parents and teachers in comparison to normative samples. The children themselves reported more than expected symptoms of depression compared to children in the general population. During interviews with parents and children these results were confirmed. Almost half of the children showed moderate to severe problems in functioning at home, at school, or in relation to peers. Until now the long-term psychosocial functioning has been a neglected area since no earlier studies relevant to this subject are available. Our study shows that there is a need for further follow-up studies of children with CDH, because serious problems in long-term psychosocial functioning may be expected.

It is concluded that children with CDH are at risk for lower cognitive and school functioning, and emotional and behavioral problems. Although the number of participants in our study was rather small, it becomes clear that children with CDH should receive an intensive follow-up to detect psychological and social problems. To clarify the issue of long-term functioning of these children a standardized follow-up program for major congenital anomalies is planned in our hospital.

Chapter 6

Long-term physical, psychological, and social functioning of children with esophageal atresia

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6.1. Abstract

Aim. Little is known about the long-term psychosocial functioning of children with esophageal atresia (EA). The physical, psychological, and social functioning of children with EA was studied using standardized assessment procedures, and relations between medical and social background variables and outcome were investigated.

Methods. Subjects were 36 children (20 boys, 16 girls; mean age 10.2) with EA. 14 children had minor and 11 children had major associated congenital anomalies. According to the 'Montreal' classification 8 children with major congenital anomalies who had also been dependent on artificial ventilation as newborns fell in the high risk class.

The children were subjected to an intelligence test. Parents completed standardized questionnaires concerning emotional and behavioral problems, psychosocial stress, and family functioning, children concerning depression and self-esteem, and teachers concerning emotional and behavioral problems. Results were compared with normative data from the general population and correlations between background and outcome variables were computed.

Results. According to Desjardins' classification 16 children had excellent, 9 children had good, and 4 children had fair outcome. The mean IQ of the children was 90.2 which is almost 10 points lower than the standardized norm of 100 ($p < .01$). High-risk children ($n = 7$) had a significantly

lower IQ (Mean IQ = 79.4, $p < .05$). Five times as many children ($n = 8$, 22 %) as in the general population (4 %) required special education ($p < .001$). More than twice as many children (30 to 35 %) as in the general population (15 %) showed elevated rates of emotional and behavioral problems as reported by parents and teachers ($p < .02$). The children themselves did not report more negative self-esteem or more depressive symptoms than children in the general population. Children with a lower IQ reported lower scholastic competence ($r = .38$, $p < .05$) and showed more emotional and behavioral problems as reported by teachers ($r = -.43$, $p < .05$). Family functioning and levels of psychosocial stress were the same as in the general population. Children in worse functioning families showed more emotional and behavioral problems as reported by parents ($r = .37$, $p < .05$) and higher depression scores as reported by themselves ($r = .47$, $p < .01$).

Conclusion. In a follow-up study using standardized assessment procedures it was shown that children with EA have more learning, emotional, and behavioral problems than children in the general population. A high-risk group of children with major associated congenital anomalies who had been ventilated as a newborn, were at special risk for cognitive problems.

6.2. Introduction

Esophageal atresia (EA) was first described in 1670 by William Durston and more elaborately by Thomas Gibson in 1697 (Myers, 1986). After the first successful operations during the forties of this century many innovations in operation techniques and management have been introduced. This has led to a survival rate of 80 to 90 % for all patients with EA (Lindahl, 1995). However, high-risk groups with mortality rates of almost 70 % can be identified (Poenaru, Laberge, Neilson, & Guttman, 1993). Follow-up studies showed that EA is associated with no or minor physical problems in the majority of cases and more serious physical problems in up to 10% of the patients (Anderson, Noblett, Belsey, & Randolph, 1992; Chetcuti, Dickens, & Phelan, 1989; Chetcuti, Myers, Phelan, Beasley, & Dickens, 1989; Chetcuti & Phelan, 1993; Lindahl, 1984; Lindahl, Rintala, & Sariola, 1993; Puntis, Ritson, Holden, & Buick, 1990; Spitz, 1992). Only a few follow-up studies are concerned with psychosocial functioning of patients with EA (Chetcuti, Myers, Phelan, & Beasley, 1988; Dera, Mies, & Martinus, 1980; Lindahl, 1984; Ure et al., 1998). Some of these studies reported normal functioning at follow-up (Chetcuti et al., 1988; Lehner, 1990; Ure et al., 1998), while other studies reported cognitive and emotional problems (Dera et al., 1980; Lindahl, 1984). A major drawback of these studies is that the study-samples differed considerably in number, age, and assessment procedures, and that hardly any standardized instruments were used.

In the present study we assessed the physical, psychological, and social functioning of children with EA using interviews and standardized instruments concerning cognitive and psychosocial functioning.

6.3. Materials and methods

6.3.1. Subjects

Thirty-six children with EA, 20 boys and 16 girls, aged 8 to 12 years (Mean age 10.2 years), treated in the pediatric surgery department of a large university children's hospital, participated in the study. Demographic and medical characteristics are given in Table 6.1. The children were classified according to the 'Montreal classification' proposed by Poenaru et al. (1993). They classified children with either life-threatening associated anomalies or with major associated anomalies who had been dependent of artificial ventilation in high-risk class II and all other children in low-risk class I. Twenty-eight children in our group were assigned to Montreal class I and 8 to Montreal class II. The nature of the associated congenital anomalies are summarized in Table 6.2.

Table 6.1
Demographic and medical characteristics

Characteristics	n
Age	8 - 12 years (Mean 10.2)
Sex Ratio	20 boys, 16 girls
<i>Socio-economic status*</i>	
Low SES	5
Middle SES	18
High SES	13
<i>Type of Esophageal atresia</i>	
EA with tracheo-esophageal fistula	31
EA without fistula	5
Duration of first hospital stay	13 - 690 day (Median 58.5 days)
Operations to follow-up	1 - 9 (Median 3)
Esophageal dilatations	0 - 32 (Median 1.5)
<i>Montreal Classification:</i>	
Low-risk class I	28
High-risk class II	8

* Socio-Economic Status based on the highest occupational level in the family (Van Westerlaak, Kropman, & Collaris, 1975).

Table 6.2

Associated congenital anomalies

Type of anomaly	n*
None	13
Minor	14
Open ductus arteriosus	3
Anomalies of larynx, trachea, and bronchi	8
Urogenital anomalies	12
Skeletal anomalies	7
Major†	11
Cardiac anomalies	6
Hydrocephalus	1
Anorectal malformations	4
Omphalocele	1
Small bowel atresia	1

* One patient can have more than one associated congenital anomaly.

† Major anomalies are life-threatening anomalies and any neurological anomaly.

6.3.2. Methods

The children and one of their parents visited the hospital for follow-up assessments. A pediatric surgeon performed a physical examination. Number of physical complaints as reported by the children and the parents was recorded. IQ of the children was tested with 2 subtests, Vocabulary and Block Design, of the Wechsler Intelligence Scale for Children (WISC-RN) which together have a correlation of .92 with full scale IQ (Van Haassen et al., 1986).

Children completed the Self-Perception Profile for Children (SPPC) a 36-item questionnaire for 8-12 year old children measuring self-esteem (Harter, 1985). The reliability and validity of the original 6 scales, General Self-Worth, Physical Appearance, Athletic Competence, Scholastic Competence, Social Acceptance, and Behavioral Conduct, were confirmed for the Dutch translation (Harter, 1986; Van Dongen, Koot, & Verhulst, 1993). The children also completed the Abbreviated Depression Questionnaire for Children (ADQC) a 9-item questionnaire screening for depression which has good internal consistency and construct validity (De Wit, 1987).

Parents completed the Child Behavior Checklist (CBCL) to assess behavioral and emotional problems of the children (Achenbach, 1991a). The CBCL contains 120 items which are scored on a 3-point scale with 0 = not true, 1 = somewhat or sometimes true, and 2 = very true or often true. A total problem score is computed by summing all 0s, 1s, and 2s. Two broad-band groupings are scored, the Internalizing grouping including withdrawn behavior, somatic complaints without physical cause, and anxious-depressed feelings, and the Externalizing

grouping including aggressive and delinquent behavior. The good reliability and validity of the CBCL were confirmed for the Dutch translation (Achenbach, 1991a; Verhulst, Van Der Ende, & Koot, 1996). The teachers of the children completed the Teacher's Report Form (TRF) which is a teacher version of the CBCL (Achenbach, 1991b). The problem behaviors are scored on the same broad-band groupings as in the CBCL. As with the CBCL, the reliability and validity of the TRF have been confirmed for the Dutch version (Achenbach, 1991b; Verhulst, Van Der Ende, & Koot, 1997).

Family functioning was assessed with the General Functioning Scale of the Family Assessment Device (FAD). The FAD is a parent-questionnaire containing 60 items, to be answered with 'yes' or 'no', which as a whole has good reliability and validity (Miller, Bishop, Epstein, & Keitner, 1985). The General Functioning Scale of the FAD contains 12 items. The General Functioning Scale has good reliability, and because in a factor analytical study it correlated highly with the first principal component of the other 48 items of the FAD, this scale can be used as a single index representing overall family functioning with higher scores indicating worse overall family functioning (Kabacoff, Miller, Bishop, Epstein, & Keitner, 1990).

Psychosocial stress in the participating families was measured with the Life-Events Questionnaire (LEQ; Berden, Althaus, & Verhulst, 1990). The LEQ is a parent-questionnaire listing 34 possible life-events which can be indicated as present or absent during the last 12 months. The number of life-events as reported on the LEQ is a reliable and valid indicator of psychosocial stress as has been shown in a large Dutch normative sample (Berden et al., 1990).

6.4. Procedure

The study was reviewed and approved by the medical ethical review board of the hospital. Children and parents received a letter in which they were informed of the purpose and procedures of the study. They were then contacted by telephone to obtain their informed consent to participate in the study. Of a total of 41 eligible children and their parents 36 participated yielding a total response of 87.8 %. No information is available on the reasons for refusal. Mothers and also fathers, if present, of all 36 children completed the questionnaires. They did this at home and were instructed to do this independently of each other. Twenty-nine children visited the hospital for psychological testing and completion of the questionnaires. The results of the questionnaires completed by the parents will be presented for all 36 children, and the results of the psychological testing and questionnaires completed by the children for those 29 children, who visited the hospital.

6.5. Statistical procedures

The results of the physical examinations are given as frequencies. In the analyses of the results of the CBCL, mean scores were computed by adding the raw scores of the fathers and mothers and dividing these by two and if no father reports were available, the raw scores of the mothers were used. For the CBCL and TRF proportions of children with scores in the normal and deviant range are given. The deviant range is defined as problem scores above the 85.5th percentile for the Total Problems, Internalizing, and Externalizing scores, i.e. the scores in the borderline and clinical range. Deviant scores defined in this way provide the strongest possible distinction between children with psychopathology versus children without (Achenbach, 1991a). This means that 14.5 % of children in the general population show elevated rates of emotional and behavioral problems. Non-parametric binomial tests were performed to assess if the observed proportions in our group were as expected. The mean total score of the ADQC, FAD, and LEQ, and mean scores on the scales of the SPPC were compared with mean scores obtained from the Dutch norm groups for these instruments using one-sample t-tests. IQ-scores were compared with the standardized norm of 100 using one-sample t-tests, and proportion of children with IQ in the deviant range (i.e. IQ below 85) was compared to the norm (15 % having an IQ below 85). Differences in scores on outcome variables between groups with different Montreal classification were assessed using analyses of variance. To investigate relations between physical and psychosocial background variables and present psychosocial functioning Pearson or Spearman rank correlations between measures of present physical functioning, family functioning, and psychosocial stress on the one hand, and measures of psychosocial functioning of the children on the other hand were computed. All significance levels were fixed at $p < .05$.

6.6. Results

6.6.1. Physical functioning

Parents ($n = 29$) reported good health for most of the children at follow-up. According to Desjardins' classification (Desjardins, Stephens, & Moes, 1964), 16 children had excellent outcomes with no feeding, swallowing or respiratory problems at all, 9 children had good outcome with slight feeding problems or respiratory problems, and 4 children had fair outcomes with moderate feeding problems and recurrent respiratory problems. Six children had physical problems caused by associated congenital anomalies which were sometimes more serious than problems directly related to EA. Four children had problems associated with anorectal malformations (fecal incontinence, constipation) and urogenital anomalies (daily catheterizations, suprapubic catheter, hypospadias, and enuresis nocturna), two had hearing problems caused by congenital anomalies, and one had deformities of the hand necessitating reconstructive surgery.

Montreal classification was not related to differences in present physical problems as measured with total physical problems and the Desjardins' classification.

6.6.2. Cognitive and psychosocial functioning

The results of the cognitive and psychosocial functioning of the children are summarized in tables 6.3 and 6.4. The mean IQ of the children was 90.2 which is 10 points lower than the standardized norm of 100 ($p < .01$). Twenty-two percent of the children ($n = 8$) required special education versus 4 % in the general population ($p < .001$) (Pijl, 1997). The mean IQ of the children in Montreal class I ($n = 7$) was 14 points lower than of the children in Montreal class II ($p < .05$)

Table 6.3

Mean IQ-scores and mean scores on questionnaires concerning psychosocial functioning vs normscores for the whole group and mean scores of Montreal class I vs Montreal class II.

Outcome	Norm Score	Esophageal Atresia N = 36	Montreal class	
			I n = 28	II n = 8
WISC-RN	100	90.3 (16)†	93.5 (13.9)	79.4 (19.6)*
CBCL Total problems	21.19	23.0 (19.7)	22.7 (18.5)	24.0 (25.6)
TRF Total problems	19.50	23.3 (22.5)	20.7 (20.7)	33.7 (28.0)
General self-worth	3.28	3.47 (.49)	3.4 (.49)	3.6 (.47)
Scholastic competence	2.80	3.01 (.67)*	3.0 (.67)	3.0 (.72)
Depression	1.73	1.34 (1.9)	1.0 (1.3)	2.6 (3.0)
Family functioning	20.1	19.0 (4.33)	18.9 (4.1)	19.0 (5.4)
Psychosocial stress	1.50	1.46 (1.5)	1.5 (1.5)	1.2 (1.6)

Note: Mean scores were compared to normscores using one sample t-tests and scores of Montreal class I were compared to Montreal class II using independent samples t-tests.

* $p < .05$, † $p < .01$

Twice as many children compared to children in the general population showed increased rates of emotional and behavioral problems as reported by parents as well as teachers. Mothers and teachers reported more internalizing problems (respectively 31 % and 37 % in the deviant range; $p < .001$), while the fathers reported more externalizing problems (29 % in the deviant range; $p < .020$). There were no differences in parent reported problem behavior between the children in the different Montreal classes. However, half of the children in Montreal class II were reported by the teachers to have elevated levels of emotional and behavioral problems, although this difference between Montreal class I and II was not significant due to the small numbers. This difference is also reflected in the fact that children with lower IQ scores showed more problem behavior at school (correlation between IQ and TRF total problem score: $r = -.45$, $p < .05$).

The children reported no higher rates of depression or lower self-esteem compared to

normative data. However, children with a lower IQ reported more feelings of depression (correlation between IQ and ADQC: $r = -.70, p < .001$) and lower school competence (correlation between IQ and the School Competence scale of the SPPC: $r = .38, p < .05$). Higher rates of present physical problems as reported by the children themselves during the physical examinations were associated with lower scores on two scales of the SPPC, i.e. General Self-Worth ($r = -.43, p < .05$) and Physical Appearance ($r = -.49, p < .01$), indicating lower self esteem in some areas for children with more physical problems. At the time of the assessment no children were treated in a mental health service.

Table 6.4

Proportion of deviant scores for IQ, CBCL, and TRF and proportion in special education for whole group and Montreal class I vs Montreal class II

Outcome	Whole	Montreal class	
	Group n = 36	I n = 28	II n = 8
IQ	30%*	26.1%	42.9%
Special Education	22%‡	21.4%	25.0%
<i>CBCL</i>			
Total problems	28.6%*	28.6%	28.6%
Internalizing	22.9%	21.4%	28.6%
Externalizing	20.0%	21.4%	14.3%
<i>TRF</i>			
Total Problems	34.5%†	30.4%	50.0%
Internalizing	37.9%‡	34.8%	50.0%
Externalizing	20.7%	13.0%	50.0%*

Note: Proportions with deviant scores were compared to proportions in general population using non-parametric binomial tests. Proportions in general population:

IQ: 15 % < 85; special education: 4 %; CBCL and TRF: 14.5 % in deviant range .

* $p < .05$, † $p < .01$, ‡ $p < .001$

6.6.3. Family functioning and psychosocial stressors

The mean total score on the FAD was similar to the mean score and standard deviation in the Dutch normative sample and level of psychosocial stress within the family in the year previous to the assessment was not higher than in the general population. Higher rates of parent- and child-reported behavioral and emotional problems were reported for children in worse functioning families (correlation between FAD and CBCL total problem score: $r = .37, p < .05$; correlation between FAD and ADQC: $r = .47, p < .01$) and for children with higher rates of psychosocial stress in the previous year (correlation between LEQ and CBCL total problem score: $r = .49, p < .01$).

6.7. Discussion

We studied the physical, intellectual, and psychosocial functioning of 36 children aged 8 to 12 years old who were born with EA. Ten percent of the children had moderate swallowing and respiratory problems directly related to the EA. Fifteen percent of the children had physical problems associated with other congenital anomalies, especially anorectal malformations. These results are comparable to other recent follow-up studies of children with EA (Anderson et al., 1992; Chetcuti, Dickens, et al., 1989; Chetcuti, Myers, et al., 1989; Chetcuti & Phelan, 1993; Lindahl, 1984; Lindahl et al., 1993; Puntis et al., 1990; Spitz, 1992). Across all studies, 90% of the children had excellent or good outcomes. Most studies focused on gastro-intestinal and respiratory problems. With the exception of the spinal and chest-wall deformities described by (Chetcuti, Dickens, et al., 1989; Chetcuti, Myers, et al., 1989) no other studies described the long-term consequences of associated anomalies such as anorectal malformations. We found that these consequences were sometimes more problematic than those directly related to EA.

This is the first follow-up study of children with EA in which the psychological and social functioning of the children was investigated using a broad range of standardized assessment procedures of parents and children. We found that children with EA had considerably lower IQ scores than children in the general population and that their participation in special education was 5 times as frequent as in the general population. Two other studies assessed IQ in children with EA (Dera et al., 1980; Lindahl et al., 1993). In the study by Dera et al. (1980) 10 children (mean age 4.3 years) with EA had a mean IQ of 91.8, which is comparable our results. Lindahl et al. (1993) reported on 33 children with EA who all had an IQ in the normal range. This result is probably strongly influenced by the fact that these children did not have associated congenital anomalies. In our group of high-risk children, who all had major associated congenital anomalies, the mean IQ was more than 20 points lower than the norm (77.6), while the mean IQ of low-risk children was in the normal range (94.8). Low IQ level was highly correlated with higher rates of self-reported depressed feelings and moderately correlated with higher rates of teacher-reported emotional and behavioral problems. Thus, it appears that the Montreal classification does not only have value in predicting mortality, but that it also presents as a prognostic indicator of long-term cognitive problems in children with EA. The conclusion seems justified that EA in itself does not cause long-term serious problems in cognitive functioning, but that these problems are related to the frequently associated major congenital anomalies.

Our study showed that children with EA had significantly more emotional and behavioral problems as reported by parents and teachers on standardized questionnaires than children in the general population. The children themselves did not report more feelings of depression or lower self-esteem. Neither presence of associated congenital anomalies nor parameters of early physical functioning such as length of the first hospitalization or the number of operations were related to present psychosocial functioning. However, children with more present physical problems showed less general self-worth and more negative perception of their physical appearance. In five

other follow-up studies of children with EA aspects of psychosocial functioning have been studied (Chetcuti et al., 1988; Dera et al., 1980; Lehner, 1990; Lindahl, 1984; Ure et al., 1998). Lindahl (1984) found that 33 children with EA had normal body image but that there were indications for increased emotional problems such as inhibition, strong self-criticism, and anxiety. Dera et al. (1980) found frequent symptoms of anxiety, regression, and disturbances of contact in 10 young children with EA. Lehner (1990) however, reported that the mental development of 122 children aged 0 to 19 years was normal and that most children visited normal schools. Chetcuti et al. (1988) and Ure et al. (1998) reported that adults with EA enjoyed normal lifestyles and normal quality of life. So, the results of these follow-up studies are conflicting. Because the samples concerned patients of very different ages, the sampling procedures varied considerably, and only a few standardized assessment instruments were used, these results are hardly comparable to the results of the present study. However, follow-up studies of children with other congenital abdominal anomalies such as diaphragmatic hernia (Bouman, Koot, Tibboel, & Hazebroek, in press) abdominal wall defects (Tarnowski, King, Green, & Ginn-Pease, 1991), and anorectal malformations (Ludman, Spitz, & Kiely, 1994; Tarnowski et al., 1991) also found increased levels of emotional and behavioral problems. So, it is plausible that psychosocial problems are not caused by any specific anomaly but that having a congenital abdominal anomaly irrespective of type is a risk factor for these problems. Possible explanations for this effect may be that parenting a child with a congenital anomaly is more difficult or that, for a child, having a congenital anomaly is a psychosocial stressor with negative consequences for its psychosocial development. An influence of medical factors such as the lengthy and frequent hospitalizations and the intensive medical procedures on later psychosocial functioning could not be shown in this study.

No other studies have addressed the question of the consequences for family functioning and levels of psychosocial stress for children with EA. We found normal family functioning and no increased levels of psychosocial stress. So, despite the enormous burden which is put on the families of these children especially in the beginning of their lives, EA seems not to lead to increased psychosocial problems within these families at long term.

It can be concluded that children born with EA who also have major associated congenital anomalies are at risk for later intellectual problems. The whole group of children with EA irrespective of associated anomalies is at risk for increased learning, emotional and behavioral problems. Few problems were reported by the children themselves as a group, but the children with a low IQ and the children with more actual physical problems showed respectively more depressive problems and a lower self-esteem in some aspects. The results of this study imply that more attention should be paid to the psychological and social functioning of individual children with EA during their development. If cognitive or psychosocial problems are recognized in time and interventions are planned, the psychosocial functioning of these children may be improved. Future follow-up studies will be needed to evaluate the course of the emotional and the behavioral problems of children with EA into adolescence and adulthood.

Chapter 7

Psychosocial functioning of children with Hirschsprung's disease or congenital anorectal malformations and their families

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7.1. Abstract

Aims of the study. Because relatively little is known of the level and determinants of the psychosocial functioning of children with Hirschsprung's disease (HD) or congenital anorectal malformations (CARM) and their families, we performed a study on this subject using standardized assessment procedures.

Methods. 50 children with HD (n = 23) or CARM (n = 27), 37 boys and 13 girls, aged 8 to 12 years were subjected to a physical examination and an intelligence test. Standardized questionnaires concerning psychosocial functioning of the children and their parents and family functioning were completed by the children, parents, or teachers.

Results. One-third of the children suffered of moderate to serious continence problems, which were more frequent in children with CARM than HD. The mean IQ of the children was 7.3 points lower than the norm of 100. Compared to normative data, twice as many children (30 %) had deviant levels of parent- and teacher- reported emotional and behavioral problems. The children did not report lower self-esteem or more depression than children in the general population. Family functioning and parental psychosocial functioning were in the normal range. No correlations were found between continence problems and self- or parent-reported psychosocial functioning of the children.

Conclusions. Children with HD or CARM are at increased risk for learning, emotional, and

behavioral problems but family and parental psychosocial functioning are in the normal range. Continence problems seem unrelated to the children's psychosocial functioning.

7.2. Introduction

Although Hirschsprung's disease (HD) and congenital anorectal malformations (CARM) are different conditions their long-term sequelae are similar. The most frequent and problematic consequences of HD and CARM are problems with fecal continence. Figures for CARM are generally worse than for HD. Very few studies are concerned with the psychosocial functioning of children with HD and CARM. Increased rates of emotional and behavioral problems and increased rates of psychiatric disturbance are reported for children and adolescents with CARM (Diseth & Emblem, 1996; Ginn-Pease et al., 1991; Ludman, Spitz, & Kiely, 1994), but not for children and adolescents with HD (Diseth, Bjørnland, Nøvik, & Emblem, 1997). Some studies found increased rates of maladjustment or worse psychosocial functioning for children with fecal incontinence versus children without (Diseth & Emblem, 1996; Diseth et al., 1997; Ginn-Pease et al., 1991), while others failed to find such a relation (Ludman et al., 1994). Family functioning and parental psychosocial functioning are very poorly studied in HD and CARM. In the one study of children with HD in which family functioning was assessed, this appeared not to be impaired in comparison to normative data (Diseth et al., 1997).

Because many questions remain concerning the problems in and determinants of the psychosocial functioning of children with HD or CARM and concerning family and parental functioning in these populations, we performed a study on the psychological and social functioning of children with HD or CARM and their families using standardized instruments. Our first aim was to assess the problems in psychosocial functioning of the children. Our second aim was to investigate relations between psychosocial functioning of the children and their families on the one hand and parameters of neonatal and present physical functioning on the other. Our third aim was to assess several aspects of psychosocial functioning of the families and to relate those to the psychosocial functioning of the children.

7.3. Materials and methods

7.3.1. Subjects

The study group consisted of children with HD or CARM who had been treated in the pediatric surgery department of a large university children's hospital in the time period from 1982 to 1986. In this time period 36 children with HD and 44 children with CARM were treated in this hospital. Eleven children (16 %), 4 with HD and 7 with CARM, died. Of the surviving 61 cases 50

children (82 %) aged 8 to 12 years, 23 with HD and 27 with CARM, and/or their parents agreed to participate in this study. Demographic and medical characteristics of the children are given in Tables 7.1 and 7.2. It is clear that both HD and CARM are serious conditions necessitating lengthy hospitalizations and many operations. The first day of hospitalization and moment of diagnosis was later for children with HD than for children with CARM. In more than 75 % of cases ($n = 21$) CARM was associated with other congenital anomalies, mainly urogenital anomalies, which was seldom the case for children with HD ($n = 1$).

Table 7.1

Demographic characteristics.

Characteristics	Hirschsprung's Disease $n = 23$	Congenital Anorectal Malformations $n = 27$
Age	8.5 - 12.0 years (Mean 10.5 years)	8.2 - 12.5 (Mean 10.6 years)
Sex-ratio	17 boys, 6 girls	20 boys, 7 girls
<i>Living arrangement</i>		
With both parents	17	23
With one parent, parents divorced	5	4
In residential care	1	0
<i>Socio-Economic Status*</i>		
Low SES	6	5
Middle SES	9	10
High SES	8	12

* Based on the highest occupational level in the family
(Van Westerlaak, Kropman, & Collaris, 1975).

7.3.2. Methods

The children and one of their parents visited the hospital for follow-up assessments. A pediatric surgeon performed a general physical examination of the children and recorded the number of physical symptoms. Specific questions concerning continence problems were asked of both children and parents, regarding: 1. the presence and seriousness of unclean underwear, 2. the presence and seriousness of constipation, and 3. the use of and problems with enemas. Each of these questions could be scored on a 4-point scale with 0 = no problems, 1 = slight problems, 2 = moderate problems, and 3 = serious problems.

Table 7.2

Medical characteristics.

Characteristics	Hirschprung's Disease n = 23	Congenital Anorectal Malformations n = 27
First day of hospitalization	1 - 455 (4*)	1- 3 (1)†
Hospital stay during first year	0 - 259 days (53)	13 - 330 days (35)
Hospitalizations to follow-up	1 - 13 (4)	1 - 25 (5)
Operations to follow-up	1- 7 (3)	1 - 7 (3)
<i>Associated Congenital Anomalies</i>		
None	22	6†
<i>Minor‡:</i>	0	13§
Urological	0	16
Genital	0	9
Skeletal	0	5
<i>Major</i>	1	8
Esophageal Atresia	0	4
Intestinal Atresia	0	1
Omphalocele	0	1
Neurological	0	3
Chromosomal	1	0

* Median in parentheses. † $p < .001$.

‡ Major associated congenital anomalies are life threatening anomalies and any neurological or chromosomal anomaly. All other anomalies are classified as minor.

§ One child may have more than one congenital anomaly.

Intelligence of the children was tested with 2 subtests, Vocabulary and Block Design, of the Wechsler Intelligence Scale for Children (WISC-RN) which together have a correlation of .92 with full scale IQ (Van Haasen et al., 1986).

Self-esteem and depression were assessed with the Self-Perception Profile for Children (SPPC) and the Abbreviated Depression Questionnaire for Children (ADQC). The SPPC is a 36-item questionnaire for 8-12 year old children measuring self-esteem. The reliability and validity of the original 6 scales, General Self-Worth, Physical Appearance, Athletic Competence, Scholastic Competence, Social Acceptance, and Behavioral Conduct, were confirmed for the Dutch translation (Van Dongen, Koot, & Verhulst, 1993). Higher scores on the SPPC indicate better self-esteem. The ADQC is a 9-item questionnaire screening for depression in children which has good internal consistency and construct validity, with higher scores indicating worse depression (De Wit, 1987).

Behavioral and emotional problems of the children were assessed with the Child Behavior Checklist (CBCL) and Teacher's Report Form (TRF). The CBCL is a parent questionnaire containing 120 items which are scored on a 3-point scale with 0 = not true, 1 = somewhat or sometimes true, and 2 = very true or often true. A total problem score is computed by summing

all 0s, 1s, and 2s. Two broad-band groupings are scored, the Internalizing grouping including withdrawn behavior, somatic complaints without known physical cause, and anxious-depressed feelings, and the Externalizing grouping including aggressive and delinquent behavior. The good reliability and validity of the CBCL were confirmed for the Dutch translation (Verhulst, Van Der Ende, & Koot, 1996). The TRF is a teacher version of the CBCL which is scored in an identical way. As with the CBCL, the reliability and validity of the TRF have been confirmed for the Dutch version (Verhulst, Van Der Ende, & Koot, 1997).

Family functioning was assessed with the General Functioning Scale of the Family Assessment Device (FAD). The FAD is a parent-questionnaire containing 60 items, to be answered with 'yes' or 'no', which as a whole has good reliability and validity (Miller, Bishop, Epstein, & Keitner, 1985). The General Functioning Scale of the FAD contains 12 items. The General Functioning Scale has good reliability, and because in a factor analytical study it correlated highly with the first principal component of the other 48 items of the FAD, this scale can be used as a single index representing overall family functioning with higher scores indicating worse overall family functioning (Kabacoff, Miller, Bishop, Epstein, & Keitner, 1990).

Parental psychosocial functioning was assessed with questionnaires concerning psychopathology and marital relationship. Psychopathology of the parents was measured with the 12-item version of the General Health Questionnaire (GHQ-12). The GHQ-12 is a measure for non-psychotic psychological problems, especially emotional problems such as depression and anxiety. The reliability and validity of the GHQ-12 are good (Koeter & Ormel, 1991). Higher scores on the GHQ indicate increased psychopathology. As a measure for the marital relationship the Interactional Problem Solving Questionnaire (IPSQ) was used. The IPSQ measures the problem solving abilities of partners in a marital relationship and can be considered as a one-dimensional measure of the quality of the marital relationship. Based on the raw scores, decile scores for the father, mother, and couple based on normative data can be computed. The internal consistency and the construct validity of the IPSQ are satisfactory (Lange, 1983).

Psychosocial stress in the participating families was measured with the Life-Events Questionnaire (LEQ). The LEQ is a parent questionnaire listing 34 possible life-events which can be indicated as present or absent during the last 12 months. The number of life-events as reported on the LEQ is a reliable and valid indicator of psychosocial stress as has been shown in a large Dutch normative sample (Berden, Althaus, & Verhulst, 1990).

7.4. Procedure

The study was reviewed and approved by the medical ethical review board of the hospital. The children and their parents received a letter in which they were informed of the purpose and procedures of the study. They were then contacted by telephone to ask their participation and to obtain their informed consent. The mothers, and if present the fathers, of all 50 children

completed the CBCL, FAD, LEQ, GHQ, and IPSQ. They did this at home, but were instructed to complete the questionnaires independently. Thirty-eight children (15 HD and 23 CARM) visited the hospital for the physical examination, psychological testing, and completion of the SPPC and ACDQ. The teachers of the children were sent the TRF. The results of the questionnaires will be presented for all 50 children, and the results of the physical examinations and IQ tests for the 38 children, who were interviewed and tested.

7.5. Statistical procedures

The results of the physical examinations are given as frequencies. A total 'problems with continence' score was computed by adding the scores on the 3 relevant variables (unclean underwear, constipation, enemas). Because there were high Pearson correlations between the parents' and children's scores on the variables 'problems with continence' ($r = .75$) and 'unclean underwear' ($r = .59$), these scores were combined by adding their scores and dividing them by two or giving the parents' or children's score if either one was missing. In the analyses the total scores of these variables were used if appropriate (e.g. to compute correlations). These combined variables were also recoded in a 3-point scale with 0 = no problems reported by both parents and children, 1 = minor problems reported by either parents or children, and 2 = moderate to serious problems reported by either parents or children.

CBCL mean scores were computed by adding the raw scores of the fathers and mothers and dividing these by two. If no father reports were available, the raw scores of the mothers were used. For the CBCL and TRF proportions of children with scores in the normal and deviant range are given. The deviant range is defined as problem scores above the 85.5th percentile for the Total Problems, Internalizing, and Externalizing scores, i.e. the scores in the borderline and clinical range. Deviant scores defined in this way provide the strongest possible distinction between children with psychopathology versus children without (Verhulst et al., 1996). Thus, 14.5 % of children in the general population have elevated rates of emotional and behavioral problems. Non-parametric binomial tests were performed to assess if the observed proportions in our group were as expected. The mean total score of the ADQC, FAD, GHQ, and LEQ, and mean scores on the 6 scales of the SPPC were compared with mean scores obtained from the Dutch norm groups for these instruments using one-sample t-tests. The proportion of parents with scores in the lowest 2 deciles of the IPSQ, representing the lowest quality of the marital relationship, was compared to normative scores. IQ-scores were compared with the standardized norm of 100 using one-sample t-tests, and proportion of children with IQ in the deviant range (i.e. IQ below 85) was compared to the norm (15 % IQ below 85). Differences in mean scores and in observed proportions in the deviant range of outcome variables between children with HD or CARM were assessed using analyses of variance and χ^2 tests. Pearson or Spearman rank correlations were computed between measures of past and present physical functioning (i.e. number of life-time

operations and hospitalizations, presence and seriousness of congenital malformations, number of present physical complaints, and seriousness of continence problems and unclean underwear) and measures of family functioning and psychosocial stress on the one hand, and measures of psychosocial functioning of the children and the parents on the other hand were computed. All significance levels were fixed at $p < .05$.

7.6. Results

7.6.1. Physical functioning

The general health of almost all children who were examined was good. One child with HD had some complaints of headache. Of the children with CARM one child had some respiratory complaints due to asthma, one child had complaints of backache, and one other had complaints about pain in his leg after a fracture. All these complaints were minor and no child had functional limitations. Four children with CARM also had esophageal atresia. Three of these had some problems with swallowing. Fourteen children had associated urogenital anomalies. One of these had a suprapubic catheter which did not give serious problems for the parents or the child, although she was very reluctant to learn self-catheterization. One child had moderate enuresis nocturna, and one had a narrowing of the urethra which needed treatment. As is shown in Table 7.3 children with CARM had significantly more continence problems than children with HD. Children with associated urogenital anomalies had much more continence problems than children with CARM alone or children with HD.

Table 7.3

Continence problems for children with HD versus CARM and for children with and without associated urogenital malformations.

Continence problems present	Hirschsprung's Disease n = 16	Congenital Anorectal Malformations n = 23	Urogenital anomalies absent n = 25	Urogenital anomalies present n = 14
<i>Unclean underwear</i>				
Absent	9	6	14	2
Minor	7	8	11	3
Moderate to serious	0	9†	0	9‡
<i>Total continence problems</i>				
Absent	7	6	12	1
Minor	5	4	8	1
Moderate to serious	4	13*	5	12‡

* $p < .05$. † $p < .01$. ‡ $p < .001$

7.6.2. Psychological and social functioning

The results of the intelligence test and the questionnaires concerning psychosocial functioning are presented in Table 7.4. The mean IQ of the children was 92.3 which is 7.7 points lower than the standardized norm of 100 ($p < .01$). Although the mean IQ of the children with CARM was somewhat higher (93.2) than that of the children with HD (90.9), this difference was not significant. The mean IQ of the children with major associated anomalies ($n = 5$) was even lower (83.4) but this difference was not significant due to the small number of children in this group. Eighteen percent of the children required special education, which is 4.5 times as many ($p < .001$) as the 4 % participation in special education in the general population (Pijl, 1997). There was no significant correlation between medical parameters such as length or number of hospitalizations, and number of operations and IQ.

Table 7.4

Mean scores of study group vs normscores and proportions of scores in the deviant range on IQ and questionnaires on psychosocial functioning, and participation in special education for the study group vs normproportions.

Outcome	Norm Score M	Study Group M (SD)	Norm Proportion %	Study Group %
IQ	100	92.3 (16.2)†	15	23.7
Special education	-	-	4	18.0‡
Problem behavior				
<i>Parents</i>				
Internalizing	5.35	6.61 (4.8)	14.5	27.1*
Externalizing	7.47	7.79 (5.8)	14.5	12.5
Total problems	21.03	24.56 (13.7)	14.5	33.3‡
<i>Teachers</i>				
Internalizing	5.80	7.12 (6.7)	14.5	30.0†
Externalizing	6.34	8.67 (11.4)	14.5	27.5*
Total Problems	21.72	28.60* (20.8)	14.5	30.0†
Self-esteem	3.28	3.41 (.44)	-	-
Depression	1.73	1.27 (1.5)	-	-

* $p < .05$, † $p < .01$, ‡ $p < .001$

The mean scores on the questionnaires were not higher than the norm scores, except for the higher mean total problem score on the TRF compared to the mean score in the general population. However, more children than expected showed deviant scores on the CBCL and TRF. They showed especially high rates of internalizing problems. IQ of the children was not related to problem behavior, but the mean CBCL total problem score and externalizing score of children from special schools ($n = 7$) were significantly higher compared to children from regular schools

($n = 41$) (respectively: total problem score 38.3 vs 22.2, $p < .01$; externalizing score 12.1 vs 7.0, $p < .05$). CBCL or TRF scores were not related to diagnosis, HD versus CARM, or to other medical characteristics. Neither were there any significant correlations between present physical functioning and CBCL or TRF total problem scores. The presence or seriousness of continence problems (unclean underwear and total continence problems) was not related to emotional or behavioral problems as reported by parents or teachers.

The children with HD and CARM did not report more symptoms of depression or lower self-esteem than children in the general population. There were no differences between diagnostic groups or in relation to other medical characteristics. Continence problems were not related to the scores on the SPPC scales or the total depression score of the children.

7.6.3. Family and parental functioning

The results of the questionnaires concerning family functioning, parental psychopathology, marital relationship, and psychosocial stress are summarized in Table 7.5. There were no differences concerning these parameters between the families and parents of the children with HD or CARM compared to the general population. There were no differences in family functioning, parental psychopathology, marital relationship, and psychosocial stress between diagnostic groups or between families or parents of children with versus those without continence problems. Problem behavior was related to family functioning as can be concluded from a moderate correlation between family functioning and the CBCL total problem score ($r = .43$, $p < .01$) and between family functioning and the CBCL externalizing score ($r = .42$, $p < .01$).

Table 7.5.

Mean scores of study group on parental questionnaires concerning family functioning, psychosocial stress and parental psychopathology vs norm scores, and proportion of scores in lowest 2 deciles for marital relationship of both parents vs normproportion.

Outcome	Norm score	Study Group
	M	($n = 50$) M (SD)
Family functioning	20.9	19.1 (3.9)
Psychosocial stress	1.68	1.71 (1.8)
<i>Parental psychopathology</i>		
Mothers	1.51	1.04 (2.1)
Fathers	1.30	1.02 (2.5)
<i>Marital relationship</i>		
Mothers	20 %	6.8 %
Fathers	20 %	4.7 %

7.7. Discussion

We studied the physical, psychological, and social functioning of 50 children with HD or CARM. The general health of these children was good. No children had serious functional limitations. In our study it was confirmed that continence problems are a frequent and serious problem for children with CARM but to a much lesser extent so for children with HD. Almost all children with associated urogenital malformations had moderate to serious continence problems. This is an indication that for those children with more complex and extended anomalies successful operation and more or less normal bowel function is hardest to achieve. Our results are comparable to the results for HD as well as CARM reported in the literature (Diseth & Emblem, 1996, Diseth et al., 1997, Ginn-Pease et al., 1991; Ludman et al., 1994). Although the concurrence of CARM and urogenital malformations is well-known very few studies paid attention to the negative effect of these associated anomalies on fecal incontinence. Only Misra, Mushtaq, Drake, Kiely, & Spitz (1996) reported that associated urogenital anomalies are a cause of more severe functional problems.

We found that children with HD or CARM had lower cognitive functioning and that they showed much more learning problems than children in the general population. The mean IQ of children in our study was 7.7 points lower than the norm and more than 4 times as many children in this group than in the general population required special education. Those children who participated in special education showed increased problem behavior, especially externalizing problems. In a study by Ginn-Pease et al. (1991) 34 children with CARM had a mean IQ of 107.9 which is above average. There is no good explanation for the difference in IQ between that study and our results. Ginn-Pease et al. (1991) found that medical complexity influenced IQ negatively. This was confirmed in our study in the sense that the presence of associated major congenital anomalies negatively influenced IQ, although due to small numbers this relation was not significant. In a study of 58 adults with CARM, Hassink, Rieu, Brugman, & Festen (1994) found that the educational level of patients with CARM was lower than in the general population. So, our results as well as results from the literature indicate that a part of the patients with HD and CARM are at risk for cognitive and educational problems, which are related to behavioral problems and which may lead to lower professional perspectives. Still, a large part of the children had IQ's in the normal range and were functioning well at school.

Compared to normative samples the children in our study showed increased levels of emotional and behavioral problems as reported by parents and teachers. The children themselves did not report higher rates of depression or lower self-esteem than children in the general population. These results confirm results of earlier studies concerning psychosocial functioning of children with CARM. Based on CBCL data, Ludman et al. (1994) and Ginn-Pease et al. (1991) found that a relatively large proportion of children with CARM showed behavioral maladjustment. Several studies also found high rates of psychiatric disturbance in children with CARM (Diseth & Emblem, 1996; Ludman et al., 1994). In contrast to our results Ludman et al.

(1994) also found higher rates of self-reported depression. Only Diseth et al. (1997) studied the psychosocial functioning of children with HD. In contrast to our results they did not find higher rates of behavioral and emotional problems or psychiatric disturbance than in the general population. Therefore, we may conclude that children with CARM are definitely at risk for psychosocial problems, while this is less certain for children with HD.

We expected that fecal incontinence would have a negative influence on psychosocial functioning. This, however, was not confirmed by our results. We failed to find a relation between continence problems and indicators of psychosocial functioning such as parent and teacher-reported problem behavior and self-reported self-esteem or depression. Previous studies which highlighted the relation of continence problems with psychosocial functioning produced conflicting results. Similar to our study, Ludman et al. (1994) did not find a relation between incontinence and behavioral maladjustment. By contrast, Ginn-Pease et al. (1991) found a significant correlation between continence and CBCL scores, but the self-concept of incontinent children was not lower than that of normal children. Diseth et al. (1996,1997) found significant correlations between incontinence and psychosocial functioning as measured with the Children's Global Assessment Scale for adolescents with HD as well as CARM.

From this controversy an important point for discussion may be raised. Although it would seem that especially the long-term problems with incontinence and constipation which necessitate chronic and sometimes very unpleasant treatment procedures are a major cause for emotional and behavioral problems, this is not necessarily the case. It has been shown that children with other congenital abdominal anomalies with less negative long-term physical consequences such as abdominal wall defects (Ginn-Pease et al.,1991), esophageal atresia (Bouman,Koot & Hazebroek, in press), and diaphragmatic hernia (Bouman, Koot,Tibboel, & Hazebroek, in press) are also at risk for increased emotional and behavioral problems. So, having a congenital abdominal anomaly appears to be a risk-factor for psychosocial problems irrespective of type of anomaly or specific long-term physical problems. There are several aspects in the development of a child with a congenital anomaly which are unrelated to the type of anomaly or long-term physical problems but may still negatively influence its psychosocial development. At the same time these are hard to grasp in a retrospective follow-up study. Problems in the attachment to the parents due to the reactions of parents to a child with a congenital anomaly or the lengthy hospitalizations in the newborn period, and interference in the emotional development due to frequent hospitalizations or painful medical procedures may all be factors related to long-term emotional and social problems. However, the effect of these factors may be obscured because there are many intermediary factors in the course of the development of a child.

Another explanation for the fact that we did not find a relation between fecal incontinence and psychosocial functioning of the children, may be that our study group consisted of school-aged children. Next to denying and being secretive about continence problems, which are frequently used coping mechanisms by 8 to 12 year-old children with continence problems (Ludman & Spitz, 1996), they may also deny their emotional problems. When the children in our

study group reach adolescence there may be a change in their psychosocial functioning related to their continence problems. Ludman and Spitz (1996) recently reported that there appears to be a shift in coping strategies with fecal incontinence during adolescence. Especially adolescent boys appear to give up their sometimes extreme denial they showed in earlier years. Although this may be accompanied by considerable stress during their adolescent years, it may eventually lead to a better acceptance of their disability. Denial is less used as a coping mechanism by school-aged girls, but they are described as very secretive about their continence problems, which continues into adolescence. Still, girls appear to come to terms with their disability at an earlier age than boys. As a consequence, it is reasonable to expect changes in the coping mechanisms of the children in our study group during adolescence which may be related to changes in their psychosocial functioning especially concerning self-esteem and relations to peers. Follow-up of our study group into adolescence is needed to clarify this issue further.

Neither family functioning nor parental psychosocial functioning and levels of psychosocial stress in the group of children with HD or CARM were more problematic than of children in the general population. Continence problems were not related to family functioning or parental psychosocial functioning. One other study assessed family functioning in children with HD (Diseth et al.1997). This study neither found more family problems in their study group. No studies are available on family functioning of children with CARM. So, although the burden of having a child with HD or CARM is high, especially if this child has sometimes severe continence problems needing daily care, parents and the families of children with HD or CARM appear to cope well. However, we found that children from worse functioning families had more behavioral problems. These results are found in many studies which relate the psychosocial functioning of children with social-ecological background variables such as family functioning (Wallander & Thompson, 1995) and is an indication of the importance of the interaction between a child and its family.

We conclude that children with HD and CARM are at increased risk for cognitive and learning problems and behavioral and emotional problems. Although many children are functioning very well there is a substantial subgroup of some 18 % with educational problems and 30 % with emotional and behavioral problems. Individual follow-up and counseling of these children and their parents is needed to help them cope with the consequences of a major congenital anomaly. Continence problems are especially frequent in children with CARM and associated urogenital anomalies, but in our group we did not detect a relation between continence problems and psychosocial functioning of the children or the parents. Longer follow-up periods are needed to see how these children develop into adolescence and which are determinants of future emotional or behavioral problems.

Chapter 8

Psychosocial adjustment of children with congenital abdominal anomalies and their families

Nico H. Bouman & Hans M. Koot

8.1 Introduction

Children with congenital anomalies form a considerable proportion of children treated in pediatric hospitals. Fifteen to twenty percent of pediatric hospital admissions concern children with congenital anomalies (Epstein, 1996). The rate of these anomalies may vary in different countries and estimates depend of the way data are collected. For western countries an estimate of 1 – 2 % of livebirth children seems reasonable (Dorrepaal, Den Ouden, & Cornel, 1998; Platt & Pharoah, 1996). The most frequent anomalies are deformities of the feet, hypospadias, cleft lip, and Down syndrome (Platt & Pharoah, 1996). Although congenital abdominal anomalies (CAA) such as diaphragmatic hernia, esophageal atresia, or anorectal malformations are relatively infrequent (incidence of 1 to 2 in 1000 live births [Beasley, Myers, & Auldist, 1991; Halsband & Von Schwabe, 1989; Langham et al., 1996; Rescorla, Morrison, Engles, West, & Grosfeld, 1992]), this is an important group in relation to amount of morbidity and quantity of medical care. Children with CAA often need long-term and intensive medical treatment, especially in the neonatal period. The mortality of congenital anomalies in general and abdominal anomalies in particular has decreased considerably during the last decades due to medical progress in surgical techniques and life-supporting technology (Halsband & Von Schwabe, 1989; Spitz, 1993). Due to these developments there exists an increasing interest in the psychosocial aspects of the prognosis of children with acquired as well as congenital medical conditions. It has been recognized that mortality and morbidity are too narrow outcome measures, and that the scope of outcome research should be enlarged to the psychological and social functioning of these children (Pantel & Lewis, 1997; Rosenbaum, Cadman, & Kirpalani, 1990).

Due to these developments there exists a growing body of research on the follow-up of children with congenital and acquired medical conditions and their families. But the need for a theoretical framework to guide research in this field has been stressed by several authors (Eiser,

1993; Wallander, Varni, Babani, Banis, & Wilcox, 1989). This framework is especially needed because the relations between medical and psychosocial factors on the one hand and outcome on the other hand is complex and cannot be analyzed in a simple linear model. Wallander and Varni (Wallander et al., 1989; Wallander & Varni, 1992; Wallander & Thompson, 1995; Wallander & Varni, 1998) proposed a disability-stress-coping model of childhood adjustment to physical disorders. In this model several factors concerning the disease or disability, psychosocial stress, stable personal factors, and social-ecological factors are incorporated and brought into relation with psychosocial adjustment. Wallander and Varni identified several risk-factors for psychosocial maladjustment: condition parameters, functional independence, and psychosocial stress, consisting of major life-events as well as disease-related problems. On the other hand they identified resistance factors which play an important role as protective factors for psychosocial maladjustment to childhood physical disorders. These resistance factors are intrapersonal factors such as competence or coping style and social-ecological factors such as family functioning, parental psychosocial adjustment, or family social support. Support for the validity of several aspects of this model has been found in the own research of the authors as well as studies of others (Wallander & Varni, 1992; Wallander & Thompson, 1995; Wallander & Varni, 1998). Wallander and Varni emphasize the generic character of this model which makes it applicable to a broad range of physical disorders. They stress the need for further research in order to come to a confirmation or adjustment of the model.

With the exception of anorectal malformations, the number of studies in which the psychosocial adjustment of children and their families with CAA was assessed in a reliable way is very limited. There hardly exist any studies in which psychosocial adjustment of these children has been brought in relation to medical as well as environmental parameters as was proposed in the Wallander-Varni model. We have used the Wallander-Varni model as a heuristic tool to generate questions and hypotheses in the study of children with CAA.

8.1.1. Hypotheses

Our first hypotheses concern the physical and psychosocial functioning of the children and their families compared to the general population.

1. *Children with congenital abdominal anomalies are at risk for problems in physical, cognitive, and psychosocial functioning.*

A wealth of studies have shown that children with a great diversity of medical conditions are at risk for increased psychosocial maladjustment (for overviews see: Eiser, 1993; Lavigne & Faier Routman, 1992; Wallander & Varni, 1998). A few studies supported this hypothesis concerning children with congenital abdominal anomalies (Dera, Mies, & Martinus, 1980; Diseth & Emblem, 1996; Ginn-Pease et al., 1991; Ludman, Spitz, & Kiely, 1994).

2. *Family functioning and parental psychosocial functioning are expected to be in the normal range compared to the general population.*

Most studies on family functioning in pediatric populations found it to be in the normal range in (see for example Cadman, Rosenbaum, Boyle, & Offord, 1991), although the number of empirically based studies on overall family functioning in childhood physical disorders is limited (Wallander & Varni, 1998).

Our second set of hypotheses concern the relations between psychosocial adjustment and the risk and resistance factors as have been postulated in the disability-stress-coping model of Wallander and Varni.

3. *Condition parameters such as diagnosis, severity or brain involvement, influence psychosocial adjustment.*

Recent literature reviews indicated that there appears to be very little effect of type or severity of disease on psychosocial functioning of children (Lavigne & Faier-Routman, 1992; Wallander & Varni, 1998). Some but not all results of several studies indicated that the psychosocial adjustment of children with Hirschsprung's disease or anorectal malformations is negatively influenced by fecal incontinence which is often present in these anomalies (Diseth & Emblem, 1996; Diseth, Bjornland, Novik, & Emblem, 1997; Ginn-Pease et al., 1991) while other results within these studies and one other study did not confirm these results (Ludman et al., 1994). However, based on clinical experience within the hospital and supported by a part of the research we hypothesized that children with Hirschsprung's disease or anorectal malformations would be at increased risk for psychosocial maladjustment.

4. *Condition parameters, such as diagnosis or severity, increase psychosocial stress.*

Conditions such as Hirschsprung's disease or anorectal malformations necessitate continuing medical treatment such as enemas or the use of laxatives which add significantly to the burden for parents and children. For these children we expected higher rates of disease-related psychosocial stress compared to children with other anomalies.

5. *Intrapersonal factors such as temperament or competence influence psychosocial adjustment.*

We considered intelligence to be an intrapersonal factor, designating competence, and we expected that higher intelligence would positively influence psychosocial functioning.

6. *Higher levels of psychosocial stress are negatively related to psychosocial adjustment.*

We expected that number of major life-events and disease-related problems as reported by children as well as parents would be related to psychosocial functioning.

7. *Social-ecological factors are related to psychosocial adjustment.*

We expected that several social-ecological factors such as overall family functioning but also the quality of the marital relationship, and parental psychopathology would influence psychosocial functioning.

The hypotheses 5 to 7 are based on the evidence provided by Wallander and Varni to support their model.

To generate answers to these hypotheses, we performed a follow-up study of a large group of unselected children with CAA using a broad range of standardized assessment instruments covering the physical, psychological, and social functioning of the children and their families.

8.2. Methods

The methods used in this study will be presented in relation to the factors incorporated in the disability-stress-coping model of Wallander and Varni.

8.2.1 Subjects

The sample consisted of 139 children, 87 boys and 52 girls, who were born in the period from 1982 to 1986 and were treated for a CAA in a large university children's hospital. A total of 226 children with CAA were treated in the hospital in the indicated time-period, of which 166 children survived. Of these 166 children, 115 children and the parents of 139 children consented to participate leaving a total response of 83.7 %. The mean age of the study group children at the time of the assessment was 10.6 years (SD 1.1).

The diagnostic categories of the children were as follows: 31 children had an esophageal atresia, 11 children a congenital diaphragmatic hernia, 19 children an abdominal wall defect, 26 children an intestinal atresia, 23 children Hirschsprung's disease, 22 children anorectal malformations, and 7 children multiple diagnoses. Most children were hospitalized in the first 3 days of their life. The duration of the first hospitalization ranged from 4 to 690 days (Median 34 days). The children were rehospitalized during their lives between 0 en 29 times (Median 3 hospitalizations) and operated between 0 en 8 times (Median 2 operations). Seventy-seven children (55 %) had only one congenital anomaly, 37 (27 %) had one or more minor associated congenital anomalies such as skeletal or urogenital anomalies, and 25 (18 %) had one or more major associated congenital anomalies such as major cardiac or neurological anomalies.

One hundred and nineteen children lived with both parents, 19 children of divorced parents lived with one parent, and 1 child was institutionalized. The distribution of family socio-economic status (SES), based on the highest parental occupational level (Van Westerlaak, Kropman, & Collaris, 1975) was: low SES n=25; middle SES n=62; and high SES n=52.

8.2.2. Measures

8.2.2.1 *Physical functioning*

The children in the study group and one of their parents were seen in the hospital for a physical examination of the child performed by a pediatric surgeon. Based on the physical examination, problems in physical functioning, i.e. neurological, cardiac, respiratory, abdominal, uro-genital, or orthopedic functioning, were scored as 0 = absent, 1 = light, 2 = moderate, or 3 = severe. A total score was computed by adding these scores. Based on the physical examinations the functional independence of the children was assessed.

8.2.2.2. *Cognitive functioning*

Intelligence was tested with two subtests, vocabulary and block design, of the *Wechsler Intelligence Scale for Children-Revised* edition, which together have a correlation of .92 with full scale IQ (Van Haassen et al., 1986). Data on participation in special education and school-level were obtained from the parents.

8.2.2.3. *Psychosocial adjustment*

Self-reports. A 'psychosocial interview' of the children concerning general psychosocial functioning was performed. This interview contained items concerning the functioning of the child within the family, at school, and in relation to peers. Problems in these areas were scored as 0 = absent, 1 = light, 2 = moderate, or 3 = severe and based on these scores a total score was computed.

Self-esteem and depression were assessed with the *Self-Perception Profile for Children* (SPPC; Harter, 1985) and the *Abbreviated Depression Questionnaire for Children* (ADQC; De Wit, 1987). The SPPC contains 36 4-point items for 8- to 12-year-old children concerning their self-esteem, and contains six subscales: Scholastic Competence, Social Acceptance, Athletic Competence, Physical Appearance, Behavioral Conduct, and General Self-Worth. In a cross-cultural validation study among Dutch children by Van Dongen, Koot, and Verhulst (1993) the factor structure and other psychometric properties presented by Harter were confirmed. The ADQC is a questionnaire with 9 2-point items which screens for depression. The ADQC has good internal consistency and construct validity (De Wit, 1987).

Parent reports. A psychosocial interview was conducted with the parents containing comparable questions to the interview with the children, yielding a total psychosocial functioning score.

Behavioral and emotional problems of the children were measured with the *Child Behavior Checklist* (CBCL, Achenbach, 1991a). The CBCL includes a competence and a problem section. The problem section of the CBCL contains 120 items concerning problem behavior which are scored on a 3-point scale with higher scores indicating more problems. A total problem score can be computed by summing the scores on these items. Eight syndrome scales

were constructed. These are Withdrawn, Somatic Complaints, and Anxious-Depressed, which together constitute the Internalizing broad-band grouping, Delinquent Behavior and Aggressive Behavior, which together constitute the Externalizing broad-band grouping, and Social Problems, Thought Problems, and Attention Problems. The competence scales of the CBCL reflect school functioning and social competence of children. A School, Social, and Activities Competence score, and a Total Competence score can be computed with higher scores indicating better functioning. The CBCL has good psychometric properties (Achenbach 1991a), which were confirmed for the Dutch translation (Verhulst, Van Der Ende, & Koot, 1996). Deviant scores on the CBCL are defined as scores above the 84.5th percentile on the Internalizing and Externalizing broad-band groupings and Total Problem scores and above the 95.5th percentile on the syndrome scales, scores below the 14.5th percentile on the Total Competence score of the CBCL, and below the 4.5th percentile on the CBCL Activities, School, and Social Competence scales. Deviant scores defined in this way provide a clear distinction between children with versus children without psychopathology (Achenbach, McConaughy, & Howell, 1987; Verhulst et al., 1996).

Teacher reports. The teachers completed the *Teacher's Report Form* (TRF, Achenbach, 1991b) The TRF is a teacher version of the CBCL. The problem behaviors are scored on the same syndromes and broad-band groupings. The competence scales of the TRF are Academic Performance and Total Competence comprising items such as attitudes toward learning, appropriate behavior, and happiness. As with the CBCL, the reliability and validity of the TRF have been confirmed for the Dutch version (Achenbach, 1999b; Verhulst, Van Der Ende, & Koot, 1997). Deviant scores on the TRF problem scales are defined in the same way as with the CBCL. Deviant scores on the TRF Total Competence score and Academic Performance score are defined as scores below the 14.5th percentile.

8.2.2.4. Risks and resistance factors

Disease parameters. From the medical charts of the children several parameters were recorded: diagnostic category, presence of major or minor associated anomalies, duration of the first hospitalization, number of rehospitalizations, and number of operations to follow-up. Problems in physical functioning were assessed during the physical examination as described above.

Psychosocial stressors. Number of major life-events in the participating families was measured with the *Life-Events Questionnaire* (LEQ; Berden, Althaus, & Verhulst, 1990). The LEQ is a parent questionnaire listing 34 possible life-events which can be indicated as present or absent during the last 12 months. The number of life-events as reported on the LEQ is a reliable and valid indicator of psychosocial stress as has been shown in a large Dutch normative sample. In addition, *disease-related psychosocial stress* was assessed. This was done by interviewing the

parents and children on those problems in the three main domains of functioning, family, school, and peers, which were specifically related to the congenital anomalies. These problems were scored as 0 = absent, 1 = light, 2 = moderate, or 3 = severe. A total 'disease-related stress score' was computed by adding the scores of these items.

Social-ecological factors. Family functioning was assessed with the *General Functioning Scale of the Family Assessment Device* (FAD; Miller, Bishop, Epstein, & Keitner, 1985). The FAD is a parent-completed questionnaire containing 60 items, to be answered with 'yes' or 'no', which as a whole has good reliability and validity (Miller et al., 1985). The General Functioning Scale of the FAD contains 12 items and has good reliability. Because in a factor analytical study it correlated highly with the first principal component of the other 48 items of the FAD, this scale can be used as a single index representing overall family functioning with higher scores indicating worse overall family functioning (Kabacoff, Miller, Bishop, Epstein, & Keitner, 1990).

As measure of the marital relationship the *Interactional Problem Solving Questionnaire* (IPSQ; Lange, 1983) was used. The IPSQ measures the problem solving abilities of partners in a marital relationship and can be considered as a unidimensional measure of the quality of the marital relationship. From the raw scores decile scores for the mother and father based on normative data can be computed. The internal consistency and the construct validity of the IPSQ are satisfactory (Lange, 1983).

Psychopathology of the parents was measured with the 12-item version of the *General Health Questionnaire* (GHQ-12; Koeter, & Ormel, 1991). The GHQ-12 is a measure for non-psychotic psychological problems, especially emotional problems such as depression and anxiety. The reliability and validity of the GHQ-12 are good (Koeter, & Ormel, 1991). Higher scores on the GHQ indicate increased psychopathology.

8.3. Procedure

The research protocol was reviewed and approved by the medical ethical board of the hospital. The children in the study group and their parents received a letter in which they were informed about the purpose and procedures of the study. They were contacted by telephone to ask their participation and obtain their informed consent.

The 115 participating children underwent a physical examination, were interviewed and subjected to an abbreviated intelligence test, and they completed the SPPC and ADQC and their parents were interviewed. The participating parents of 139 children completed the questionnaires at home. Questionnaires were completed by both parents of 120 children and in all other cases only by the mother. The parents were instructed to complete the questionnaires independently.

8.4. Results

8.4.1. Physical, cognitive, and psychosocial functioning

8.4.1.1 *Physical functioning*

Almost all children were in good health. Two children had moderate functional impairments due to hearing problems but for all other children functional independence was not limited. Twenty-seven children had moderate physical problems. The most frequent problems were gastrointestinal problems related to their congenital anomaly. Thirteen children with anorectal malformations and four children with Hirschsprung's disease had moderate to serious problems with fecal continence. Four children with esophageal atresia had problems with swallowing.

8.4.1.2. *Cognitive and school-functioning*

A one-sample t-test showed that the mean IQ of the children was significantly lower than the norm IQ (91.5 vs. 100, $p < .001$). Seventeen per cent of the children required special education, which is 4 times as many as the four per cent participation in special education reported for the general population (Pijl, 1997) (Binomial test: $p < .001$).

8.4.1.3. *Psychosocial adjustment*

Self-reports. One-sample t-tests showed that, compared to the general population, the children reported higher scores on several scales of the SPPC: Scholastic Competence (3.5 vs norm score = 2.81, $p < .001$); Physical Appearance (3.49 vs norm score = 3.16, $p < .001$); General Self-Worth (3.53 vs norm score = 3.28, $p < .001$). Further, they had lower scores on the depression questionnaire (ADQC total score = 1.26 vs norm score = 1.73, $p < .05$). These are both indications that the children in the study group had higher self-esteem and fewer depressive problems compared to the general population.

During the psychosocial interviews, 33 children reported mostly moderate problems. Twenty-eight children reported problems in one domain (family: $n = 14$; school: $n = 9$; peers: $n = 5$), 4 children in two domains (family and school: $n = 1$; family and peers: $n = 2$, school and peers: $n = 1$), and one child in all three domains.

Parent and teacher reports. One-sample t-tests showed that the mean scores on the CBCL and TRF were not higher than the normative scores. However, binomial tests showed that the proportions of scores in the deviant range (i.e. in the borderline or clinical range) on several problem and competence scales of the CBCL and TRF were higher than in the general population (Table 8.1). About 30 % of the children obtained deviant total problem scores on the CBCL as well as the TRF compared to 15 % in the general population. On all CBCL syndrome scales, except thought problems, and on the internalizing scale higher proportions of children than

expected had deviant scores. Higher than expected proportions of children had deviant TRF scores on the withdrawn and somatic complaints scale, on the aggressive behavior scale, and on both broad-band scales. On the competence scales, parents reported significantly higher than expected proportions of children with low school competence and low social competence. The teachers reported a higher than expected proportion of children with low academic performance, while the scores on other TRF competence scales were not lower than expected.

In the psychosocial interviews, the parents reported that 15 children had moderate to serious problems in one domain (family: $n = 1$; school [learning problems]: $n = 10$; peers: $n = 4$), 9 children in two domains (family and school: $n = 5$; family and peers: $n = 1$; school and peers: $n = 3$) and 2 children in all three domains. For the whole group the parents reported any problems for 26 children (19 %).

Table 8.1

Proportions of children in borderline and clinical range on the CBCL and TRF

	Parents		Teachers	
	% deviant score	p-value*	% deviant score	p-value
<i>Problem scales</i>				
Withdrawn	10.5	.0008	19.8	.0000
Somatic Complaints	15.7	.0000	11.9	.0001
Anxious-Depressed	13.5	.0000	6.8	ns.
Social Problems	17.2	.0000	7.6	ns
Thought problems	6.7	ns	5.9	ns.
Attention Problems	15.7	.0000	6.8	ns.
Delinquent Behavior	8.2	.0313	4.2	ns.
Aggressive Behavior	9.0	.0113	8.5	.0314
<i>Broad band scales</i>				
Internalizing	31.3	.0000	28.0	.0002
Externalizing	17.9	ns.	22.9	.0237
Total Problems	32.1	.0000	27.1	.0006
<i>Competence scales</i>				
Activities	5.2	ns.	-	
Social Competence	9.6	.0042	-	
School Competence	13.1	.0000	-	
Academic Performance	-		22.9	.0237
Total competence	23.2	.0121	17.0	ns

* Based on binomial tests with 4.5 % as expected value for the syndrome scales and the CBCL competence scales, and 14.5 % as the expected value for the broad band scales, the total problem scales, and CBCL total competence and TRF school performance scale

8.4.2. Risk and resistance factors

8.4.2.1. Psychosocial stress

A one-sample t-test showed that the number of life-events as reported by both parents (1.49) did not differ from the general population mean (1.50; Berden et al., 1990). Concerning disease-related stress, 28.3 % of the parents and 7.1 % of the children reported moderate to serious disease-related problems in at least one domain (family, school, peers).

8.4.2.2. Social-ecological factors

The results of family functioning and psychosocial functioning of the parents are given in Table 8.2.

Table 8.2

Mean scores of study group on parental questionnaires concerning family functioning, psychosocial stress and parental psychopathology vs norm scores, and proportion of scores in lowest 2 percentiles for marital relationship of both parents vs normproportion.

Outcome	Norm score M	Study Group M (SD)	p-value
<i>Family functioning</i>			
Mothers (n = 136)	19.9 (4.8)	19.9 (5.0)	ns.
Fathers (n=120)	20.7 (4.9)	19.4 (4.4)	.002
Psychosocial stress (n=139)	1.68 (1.7)	1.49 (1.55)	ns.
<i>Parental psychopathology</i>			
Mothers (n=133)	1.51	.94 (2.1)	.002
Fathers (n=120)	1.30	1.23 (2.3)	ns
<i>Marital relationship</i>			
Mothers (n=120)	20 %	6.6 %	.0002
Fathers (n=108)	20 %	10.2 %	.0076

One-sample t-tests showed that family functioning as reported by mothers was the same as in the general population (19.9 vs 19.9, ns), while fathers reported better family functioning (19.4 vs 20.7, $p < .01$). As was shown with binomial tests, a smaller proportion than in the general population of both fathers (10.2 %, $p < .01$) and mothers (6.6 %, $p < .001$) obtained scores on the IPSQ in the lowest 2 deciles, indicating that in this group parents reported a better marital relationship than parents in the general population. Finally, the level of non-psychotic psychopathology was lower for mothers (.94 vs 1.51, $p < .01$) and the same for fathers (1.23 vs 1.30, ns.) compared to the general population, as was shown with one-sample t-tests.

8.4.3. Relations between risk factors, resistance factors, and psychosocial adjustment

The results in this section are presented in the same order as the hypotheses based on the Wallander-Varni model. The Pearson correlations between the dependent and independent variables are given in Table 8.3.

8.4.3.1. *Relations between condition parameters and adjustment*

Almost all condition parameters, i.e. diagnosis, presence of associated anomalies, or seriousness of physical problems were not significantly related to self-reported or parent-reported psychosocial adjustment. There were low but significant Pearson correlations between parent-reported physical problems and the CBCL total problem score ($r = .24, p < .05$) and the self-reported depression score ($r = .27, p < .01$).

There was a strong relation between presence of major congenital anomalies and cognitive functioning. Children with major associated congenital anomalies ($n = 19$) had a mean IQ of 76.9 which is significantly lower than the mean IQ of children without ($n = 96$; $IQ = 94.4, p < .01$). These results are also reflected in the 44 % participation in special education of children with major associated congenital anomalies compared to 11.4 % of children without ($\chi^2 = 15.21, p < .001$) and 4 % of children in the general population (Pijl, 1997) (Binomial test $p < .001$). We did not find significant correlations between length of neonatal hospitalization, number of operations, or number of rehospitalizations and IQ.

8.4.3.2. *Relations between condition parameters and psychosocial stress*

Neither analyses of variance nor Pearson correlations showed any significant relation between diagnosis, presence of associated congenital anomalies, or seriousness of physical problems and levels of psychosocial stress, i.e. number of major life-events and self-reported or parent-reported disease-related stress.

8.4.3.3. *Relations between intrapersonal factors and psychosocial adjustment*

There were no significant Pearson correlations between IQ and any of the measures of psychosocial adjustment, except a low negative correlation between IQ and self-reported depression ($r = -.19, p < .05$).

8.4.3.4. *Relations between psychosocial stress and psychosocial adjustment*

General psychosocial stress as measured by the number of major life-events had a low but significant Pearson correlation with the CBCL total problem score ($r = .27, p < .01$), with general self worth ($r = -.20, P < .05$), and with depression ($r = .23, p < .05$). Parent-reported disease-related stress was highly correlated with the CBCL total problem score ($r = .51, p < .001$), and low but significantly with self-reported general self-worth and depression ($r = -.23, p < .05$ and $r = .23, p < .05$, respectively). Both parent-reported and self-reported disease-related stress were

significantly correlated with the TRF total problem score ($r = .44$, $p < .001$, and $r = .26$, $p < .01$, respectively). Further, self-reported disease-related stress was significantly related with self-reported general self-worth ($r = -.41$, $p < .001$) but not with depression or CBCL total problem scores.

Table 8.3

Pearson correlations between risk and resistance factors and psychosocial adjustment measures.

	CBCL Total problems	TRF Total problems	General Self Worth	Depression
<i>Risk factors</i>				
Physical problems-parents	.24*	ns	ns	.27†
Disease-related stress-parents	.51‡	.44‡	-.23*	.23*
Disease-related stress-children	ns	.26†	-.41‡	ns
Major life events	.27†**	ns	-.20*	.23*
<i>Resistance factors</i>				
Intelligence	ns	ns	ns	-.19*
Family functioning	.37‡	.21*	-.25†	.33‡
Marital relationship-mothers	-.29†	ns	ns	-.26*
Marital relationship-fathers	-.27†	ns	ns	ns
Psychopathology-mothers	.34‡	ns	ns	.28*
Psychopathology-fathers	ns	ns	ns	ns

Note: Number of subjects in the analyses varied from 92 to 133.

* $p < .05$, † $p < .01$, ‡ $p < .001$.

8.3.4.5. Relations between social-ecological factors and psychosocial adjustment

As shown in Table 8.3., family functioning had moderate to low correlations with all measures of psychosocial adjustment ($r = .21$ to $r = .37$). The quality of the marital relationship as reported by both mothers and fathers had low correlations with the CBCL total problem score ($r = -.29$, $p < .01$ and $r = -.27$, $p < .01$, respectively). In addition, the quality of the marital relationship and non-psychotic psychopathology as reported by the mothers had low correlations with child-reported depression ($r = -.26$, $p < .05$ and $r = .28$, $p < .05$, respectively).

8.4.3.6. Results of multiple regression analyses

Based on the correlations between risk and resistance factors and psychosocial adjustment several multiple stepwise linear regressions were performed with CBCL and TRF total problem scores, and child-reported general self-worth and depression as dependent variables to assess the independent contribution of predictor variables to the prediction of psychosocial adjustment. As predictor variables the stress factors parent-reported physical problems, general psychosocial stress, and self-reported and parent-reported disease-related stress were entered in the analyses. Further, the resistance factors intelligence and family functioning were entered in the analyses.

Family functioning was chosen out of the social-ecological resistance factors because it correlated high with marital relationship ($r = .62$, $r = .63$, for mothers and fathers respectively) and low to moderate with parental psychopathology ($r = .21$, $r = .38$, for mothers and fathers respectively), and because it was the single social-ecological variable which correlated significantly with all measures of psychosocial adjustment. The results of the regression analyses are summarized in Table 8.4. Parent-reported problem behavior was predicted by parent-reported disease-related stress (explaining 25.1 % of the variance), family functioning (10.0 % of the variance), and parent-reported physical problems (2.7 % of the variance). Teacher-reported problem behavior was predicted by parent-reported disease-related stress (17.1 % of the variance) while all other variables did not add to the explained variance in the TRF total problem score. Self-reported general self-worth was predicted by self-reported disease-related stress (13.7 % of the variance), family functioning (6.0 % of the variance). Depression was predicted by family functioning (9.4 % of the variance) and parent-reported physical problems (5.0 % of the variance), and self-reported disease-related stress (4.5 % of the variance). Over all regression analyses disease-related stress, either parent-reported or self-reported, and family functioning together explained 17.1 % to 35.1 % of the variance in parent-reported or self-reported psychosocial adjustment.

Table 8.4

Stepwise linear regression analyses on parent and teacher-reported problem behavior and self-reported general self-worth and depression.

Step	Predictor variable*	β	Cumulated R^2	R^2 Change	P-value
<i>CBCL Total Problem score</i>					
1	Disease-related stress Parents	.405	.251	.251	.000
2	Family functioning	.301	.351	.100	.000
3	Physical complaints	.167	.377	.027	.042
<i>TRF Total Problem score</i>					
1	Disease-related stress Parents	.41	.171	.171	.000
<i>General self-worth</i>					
1	Disease-related stress Children	-.371	.137	.137	.000
2	Family functioning	-.245	.198	.060	.009
<i>Depression</i>					
1	Family functioning	.262	.094	.094	.006
2	Physical complaints	.228	.144	.050	.014
3	Disease-related stress Children	.213	.189	.045	.020

* Only those predictor variable which added significantly to the variance are entered in the table.

8.5. Discussion

This is the first follow-up study of children with CAA with a large number of participants, including all major abdominal anomalies, in which a broad range of standardized instruments concerning psychosocial functioning of the children and their families was used. We postulated several hypotheses which will be discussed presently.

8.5.1. Children with CAA are at risk for increased problems in physical, cognitive, and psychosocial functioning.

This hypothesis was largely confirmed. Most children were functioning well physically and very few children had serious functional impairments. However, 26 % of the children reported mostly moderate physical complaints, which is twice as much as can be expected (Campo & Fritsch, 1994). The rates of physical complaints in this study are comparable to other studies on CAA (Ginn-Pease et al., 1991; Diseth & Emblem, 1996; Diseth et al., 1997; Lindahl, Rintala, & Sariola, 1993; IJsselstijn, Tibboel, Hop, Molenaar, & De Jongste, 1997).

The cognitive and school-functioning of the children was considerably lower than would be expected. The mean IQ was almost 10 points lower and participation in special education was four times as frequent compared to the general population. Several other studies also reported cognitive problems for children with a diversity of CAA (Davenport, Rivlin, D'Souza, & Bianchi 1992; Dera et al., 1980; Kato et al., 1993; Nobuhara, Lund, Mitchell, Kharasch, & Wilson, 1996) while other studies did not (Dera et al., 1980; Ginn-Pease et al., 1991; Lindahl, 1984; Ludman et al., 1994). In conclusion, there is conflicting evidence concerning the co-occurrence of CAA and deficits in cognitive functioning, but based on our results we conclude that researchers and clinicians should be alert for lower cognitive functioning in children with CAA.

Concerning psychosocial adjustment, twice as many children compared to the general population showed increased levels of parent- and teacher-reported emotional and behavioral problems. The parents reported lower social competence on the CBCL and in the interviews they reported that 20 % of the children had moderate to serious problems in at least one domain of psychosocial functioning. The children themselves reported lower levels of depression and higher self-esteem compared to the general population and when they reported psychosocial problems during the interviews these were mostly moderate.

Those few studies which concerned children with CAA reported comparable levels of maladjustment (Dera et al., 1980; Diseth & Emblem, 1996; Ginn-Pease et al., 1991; Ludman et al., 1994). In contrast to these results, Diseth et al. (1997) did not find increased maladjustment in children with Hirschsprung's disease. For the children's self-reports, the results obtained in other studies are comparable to ours. Children with abdominal wall defects, Hirschsprung's disease, or anorectal malformations obtained scores in the normal range on the Piers-Harris self-concept scales or the self-report version of the CBCL (Diseth and Emblem, 1996; Diseth et al.,

1997; Ginn-pease et al., 1991). It should be emphasized that the mean CBCL and TRF scores were not higher than mean normative scores. So, it can be concluded that there is much variability in the psychosocial adjustment to CAA. Although there is a subgroup of children with CAA who show signs of psychosocial maladjustment, there is also a large proportion who are doing very well. These findings are comparable to results from many other studies concerning the psychosocial adjustment to physical disorders (Wallander & Varni, 1998).

It is important to note that the children's self-reports differed from the parents' and teachers' reports. This underscores the fact that children have a different view on their own psychosocial functioning and that children's self-reports should be incorporated in studies on childhood adjustment to physical conditions.

8.5.2. Family functioning and parental psychosocial functioning are expected to be in the normal range compared to the general population.

This hypothesis was confirmed by our results. Family functioning, marital relationship of the parents, and parental psychopathology were all in the normal range. In line with these results, there is ample evidence that family functioning as a whole and quality of the marital relationship is mostly normal in families and parents of children with congenital anomalies (Cappelli, McGrath, Daniels, Manion, & Schillinger, 1994; Diseth, Bjørdal, Schultz, Stange, & Emblem, 1998; Fletcher et al., 1995; Heller, Rafman, Zvagulis, and Pless. 1985; Holmbeck et al., 1997). In the only study in which family functioning in CAA was assessed (Diseth et al., 1997), comparable results were found. Also, several studies on the psychosocial functioning of mothers of children with congenital anomalies reported normal functioning (Capelli et al., 1994; Holmbeck et al., 1997). No studies are available on the maternal adjustment of mothers of children with CAA.

So we may conclude that the social environment in which these children grow up is not clearly different from that of children in the general population and that most families have adequate abilities to cope with the stress of having a child with a congenital anomaly.

8.5.3. Condition parameters such as diagnosis, severity or brain involvement, influence psychosocial adjustment.

We expected that children with more severe CAA would show increased psychosocial maladjustment. This hypothesis was not confirmed. There was a consistent picture across almost all results that disease parameters such as diagnosis, presence of associated congenital anomalies, or seriousness of physical problems were not related to psychosocial adjustment of the children or their families. We only found that there was a low correlation between the parent-reported physical complaints and the CBCL Total Problem score and the child-reported depression scores.

We further hypothesized that children with Hirschsprung's disease or anorectal

malformations would have more psychosocial problems, because of their frequent problems with fecal incontinence. However, this hypothesis was not confirmed. There is conflicting evidence concerning the effect of fecal continence on psychosocial functioning of children. In those studies which incorporated children's self-reports, no differences were found between children with versus children without fecal incontinence on measures of problem behavior, depression, or self-esteem (Diseth & Emblem, 1996; Diseth et al., 1997; Ginn-Pease et al., 1991; Ludman et al., 1994). Worse parent-reported global psychosocial functioning and increased problem behaviors are reported by some authors (Diseth & Emblem, 1996; Diseth et al., 1997; Ginn-Pease et al., 1991) but not by others (Ludman et al., 1994). Although fecal incontinence may cause shame in children to which they often react with denial or being secretive about it (Ludman & Spitz, 1996), it does not necessarily lead to emotional or behavioral problems.

Concerning the relation between intelligence and condition parameters, it should be emphasized that in this sample associated major congenital anomalies have a considerable negative influence on intelligence. Other physical parameters, which might be expected to negatively influence IQ, such as length of neonatal hospitalization or number of operations, were not related to IQ. No other studies related intelligence to the presence of associated anomalies, but in two other studies cognitive functioning was negatively influenced by number of operations or length of the first hospitalization (Kato et al., 1993; Ludman, Spitz, & Lansdown, 1993). So, in the case of CAA prenatal as well as postnatal factors appear to be determinants of level of cognitive functioning.

8.5.4. Condition parameters, such as diagnosis or severity, increase psychosocial stress.

There were no relations between any of the condition parameters and levels of psychosocial stress, be it general psychosocial or disease-related stress. So, psychosocial stress, and specifically disease-related psychosocial stress, does not appear to be restricted to any specific diagnostic group, but is more or less equally distributed across the different diagnostic groups. For example, for some children problems related to fecal incontinence were reported, others were reported to be ashamed and embarrassed about having a congenital anomaly or having a large operation scar, while still others were reported to have decreased physical endurance so that they could not participate in sports. Apart from studies on the consequences of fecal incontinence on psychosocial adjustment in children with Hirschsprung's disease or anorectal malformations (Diseth & Emblem, 1996; Diseth et al., 1997; Ginn-Pease et al., 1991; Ludman et al., 1994), this is the first study which shows that congenital abdominal anomalies, irrespective of type of anomaly, cause significant disease-related stress.

8.5.5. Intrapersonal factors such as temperament or competence influence psychosocial adjustment.

As we found that intelligence was not related to any measure of psychosocial adjustment except a low correlation between IQ and self-reported depression, this hypothesis has to be rejected. Very few studies on congenital anomalies investigated the relation between intelligence and psychosocial adjustment. Recent reviews on the psychosocial adjustment to physical disorders neither paid attention to this aspect (Eiser, 1990; Wallander & Varni, 1998). Wallander and Varni (1998) emphasize other intrapersonal characteristics, such as temperament or coping style, as related to psychosocial adjustment. In contrast to our results, Utens et al. (1993) found a low negative correlation ($r = -.28$) between intelligence and parent-reported problem behavior in a study on the psychosocial adjustment to congenital cardiac anomalies. No studies on CAA investigated the relation between intelligence and psychosocial adjustment.

8.5.6. Higher levels of psychosocial stress are negatively related to psychosocial adjustment.

This hypothesis is clearly supported by our results. Disease-related psychosocial stress as reported by the parents or the children had a negative relation with all psychosocial adjustment measures. The proportion explained variance in the linear regression equations varied from 4.5 % to 25.1 %. This means that problems which are directly related to the congenital anomaly may play an important role in the development of emotional and behavioral problems. This effect may partly be an explanation for the high proportion of children with emotional and behavioral problems in the deviant range in this group of children. One might argue that this measure of disease-related stress is a direct reflection of general psychosocial functioning and as such not independent from emotional and behavioral problems. However, general psychosocial functioning assessed using the psychosocial interviews had only a low correlation with disease-related stress and on the other hand a high correlation with the CBCL Total Problem score. Based on these analyses, we may conclude that the disease-related stress score is to a large extent independent of general psychosocial functioning and could as such be entered as an independent variable in the linear regression analyses.

An important conclusion can be drawn from the fact that children with CAA show increased psychosocial maladjustment which is to a considerable extent influenced by disease-related psychosocial stress, while there appear to be no relations between these variables and condition parameters. Rather diverse aspects of these anomalies cause stress and maladjustment which are not always obvious to the researcher or the clinician. So, attention for psychosocial maladjustment should not be restricted to those groups of children with CAA who a priori are expected to show problems, such as children with continence problems. Specific information should be obtained from all parents and children concerning their psychosocial functioning and the stress they experience in relation to the anomaly. Based on this information children at risk

for psychosocial maladjustment might be identified and might be offered individual or family counseling.

8.5.7. Social-ecological factors are related to psychosocial adjustment.

This hypothesis was also supported by our results. Family functioning was related to parent- and teacher-reported problem behavior as well as to self-reported depression and self-worth. This finding is a replication of the results of many studies on the psychosocial functioning of children, and reflects the very important interaction of a child with its family (Dadds, 1995; Wallander & Varni, 1998). The psychosocial functioning of children with congenital anomalies has been related to aspects of family functioning such as level of conflict and control (Murch & Cohen, 1988), family cohesiveness (Lavigne, Nolan, & McLone, 1988; Varni, Rubenfeld, Talbot, & Setoguchi, 1989), and parenting stress (Krueckeberg & Kapp-Simon, 1993). In the one study on children with congenital abdominal anomalies that assessed family functioning, it was found that parental warmth was related to psychosocial adjustment (Diseth et al., 1997). It is important to stress that the relation between family functioning and psychosocial adjustment is reciprocal, which means that problematic family functioning may cause psychosocial problems in a child but also that problematic psychosocial functioning of a child may cause problems in family functioning. These findings, however, underscore the importance to assess family functioning in follow-up studies concerning psychosocial functioning of children with congenital abdominal anomalies.

8.5.8. Conclusion

The Wallander-Varni disability-stress-coping model proved to be a useful tool in order to identify important determinants of psychosocial adjustment for children with congenital abdominal anomalies. Several aspects of the model could be verified in this sample. Both disease-related psychosocial stress and family functioning had an important influence on psychosocial adjustment. Because almost none of the children had physical limitations, the effect of functional dependence on psychosocial adjustment could not be assessed. While Wallander-Varni in their model place cognitive functioning under disease parameters, we are inclined to view intelligence as an important intrapersonal factor which denotes one aspect of personal competence, i.e. cognitive competence. This aspect of intrapersonal functioning appears to be related to specific disease parameters, such as presence of major congenital anomalies (this study) or neonatal physical factors such as surgical stress or number of operations (Kato et al., 1993; Ludman et al., 1993).

We conclude that children with congenital abdominal anomalies are at risk for cognitive, learning, and emotional and behavioral problems. These problems are not related to any specific diagnosis. The families and parents of these children are mostly functioning normally indicating

that most families and parents have important coping resources which make them able to deal well with these severe congenital anomalies and their consequences. We found important relations between disease-related psychosocial stress, family functioning, and psychosocial adjustment of the children. These relations are not limited to specific anomalies which indicates that there should be an alertness for psychosocial problems and disease-related stress for all children with congenital abdominal anomalies. Follow-up programs should take these conclusions into account. Intervention programs for children with CAA should be aware of the fact that these children, especially when they have multiple major anomalies are at risk for cognitive and learning problems. School-based interventions may be needed for these children. Further, it should be recognized that two groups of children are at increased risk for psychosocial maladjustment: first, children who come from problematically functioning families, and second, children who indicate that they experience psychosocial problems within the family, at school, or in their peer group which are directly related to their anomaly. The only way to identify these children is by asking these children and their families specifically about these problems. Then, adequate interventions can be planned to offer guidance and support to these children and their families.

Chapter 9

General discussion

9.1. Aims of the study

Congenital abdominal anomalies (CAA) are life-threatening anomalies which cause considerable mortality ranging from 6 % for Hirschsprung's disease (Rescorla, Morrison, Engles, West, & Grosfeld, 1992) to 40 % for congenital diaphragmatic hernia (Langham et al., 1996), serious neonatal and in some cases long-term morbidity (Affourtit, Tibboel, Hart, Hazebroek, & Molenaar, 1989; Ditesheim & Templeton, 1987; Lindham, 1984; Puntis, Ritson, Holden, Buick, 1990; Rescorla et al., 1992). Concerns are raised by the parents and treating physicians about the future physical and psychosocial development of these children. The scope of many existing follow-up studies on CAA is too limited to give answers to these concerns. This induced the departments of child and adolescent psychiatry and pediatric surgery to perform a follow-up study in which a large non-selected group of children with CAA participated who were investigated with a broad range of instruments to assess the physical, psychological, and social functioning of the children, family functioning, and psychosocial functioning of the parents.

The aims of the study were:

1. To assess the health-related quality of life of children who are born with a CAA when they reach middle childhood.
2. To assess the outcome of children with a CAA in terms of physical, cognitive, and psychosocial functioning.
3. To investigate which are the major determinants of the outcome of children with a CAA.

This is the first study in which a large unselected group of children with all types of major congenital abdominal anomalies and their parents participated, in which the physical, cognitive, and psychosocial functioning of the children was assessed using a broad range of standardized instruments, and in which standardized instruments for family functioning and psychosocial functioning of the parents were included.

The results of this study will be discussed in relation to the aims of the study and in the given order.

9.2. Medical characteristics of the study group

The study group consisted of 139 non-selected children with a major CAA: congenital diaphragmatic hernia ($n = 11$), esophageal atresia ($n = 31$), abdominal wall defect ($n = 19$), intestinal atresia ($n = 26$), Hirschsprung's disease ($n = 23$), anorectal malformation ($n = 22$), and a group with multiple diagnoses ($n = 7$). Almost all children were hospitalized in the first three days of their lives. The median duration of the first hospitalization was 34 days (range 4 – 690 days), the median number hospital readmissions was 3 (range 0 – 29 times), and the median lifetime number of operations was 2 (range 0 – 8 times). Forty-five per cent of the children had one or more associated congenital anomalies (37 [27 %] a minor and 25 [18 %] a major anomaly). From these figures it is clear that all children had serious anomalies necessitating sometimes very lengthy and intensive medical treatment. Given these figures, the disease-related and psychosocial burden for the children and their parents must at times have been very high.

9.3. The health-related-quality of life of children with CAA

During the last decades, the interest in the quality of life of people with physical disorders is growing. Due to medical progress the possibilities to treat serious medical conditions have increased, but concerns are raised about how some treatment strategies may affect the well-being of patients (Mulhern et al., 1989). Second, it is recognized that mortality and morbidity are a too narrow scope for follow-up studies of physical disorders and that aspects of psychological and social functioning should be incorporated in outcome studies (Pantell & Lewis, 1987; Rosenbaum, Cadman, & Kirpalani, 1990). One line of research in this field can be referred to as health-related quality of life research (HRQOL). Spieth and Harris (1996) discern two conceptual models in HRQOL research: the health-utility model and the health status measurement. In the first model (Kaplan and Anderson, 1988) values are assigned to different health-states, which are used in cost-effectiveness analyses of medical treatments. Second, the health status measurement in which HRQOL is conceptualized as a multidimensional construct including the physical, psychological, and social functioning domains (Aaronson, 1991; Landgraf & Abetz, 1996; Mulhern et al., 1989; Spieth & Harris, 1996). Because the utility model may not be applicable for pediatric populations (Spieth & Harris, 1996), the health status measurement approach has been adopted in the majority of HRQOL studies in children. In this study we developed a HRQOL instrument for children which will be discussed presently.

9.3.1. Development of a health-related quality of life instrument for children

The goal of this study was to develop a HRQOL instrument for children which should measure the functioning of the children in the physical, psychological, and social functioning domains.

Based on existing literature concerning HRQOL, within each domain several subdomains were chosen. Subdomains included in the physical functioning domain were physical well-being, physical complaints, and functional status; subdomains included in the psychological functioning domain were general psychological well-being, emotions, self-esteem, and cognitive functioning; subdomains included in the social functioning domain were relations within the family and with peers, functioning at school, and leisure time activities. Based on own clinical experience, existing instruments for psychosocial functioning of children, and peer review, items were generated covering the domains and subdomains in an adequate way. As we wanted to develop an instrument which could be completed by the parents as well as the children we made a child- and parent-version. We chose a questionnaire as the format for the instrument including questions which could be answered on a 3-point scale defined as 0 = seldom or never, 1 = sometimes, and 2 = always or mostly. The questionnaire was named the Quality of Life Questionnaire for Children (QLQC). A pilot-study was performed to test the feasibility of the QLQC. After this pilot-study some items were deleted and some items were added. The QLQC used in the present study eventually contained 118 items.

The subjects included in the quality of life study were the 139 children with congenital abdominal anomalies (study group) and 136 children from the general population matched with the study group for age, gender, and degree of urbanization (comparison group). Several self-report and parent-report instruments concerning the psychosocial functioning of the children were included in the study to test the validity of the QLQC. To assess the test-retest reliability, almost sixty children completed the QLQC 2 weeks after the first time for the second time.

Exploratory and confirmatory factor analyses yielded a structure of the QLQC with 3 scales in the physical functioning domain (18 items), 4 scales in the psychological functioning domain (32 items), and 5 (parents) or 6 (children) scales in the social functioning domain (35 items). Identical scales were constructed for parents and children, except that a 'relation with parents' scale could only be confirmed for the children's version.

The reliability measures, i.e. internal consistency and test-retest correlations, of the three overall domain scales, the subscales of the psychological functioning domain, and several subscales of the social functioning domain were good. However, the reliability of the subscales in the physical functioning domain were rather weak. This is probably caused by the fact that the physical functioning of the children in the comparison group as well as the study group was relatively good. So, the variance in physical functioning was low with the effect that small inconsistencies or changes over time in the scores may have had a relatively large effect on reliability measures. Other scales with insufficient reliability were the Relations with Parents scale, the Relations with Siblings scale, and the School functioning scale in the social functioning domain. The first two scales probably contained too few items (3 and 4 respectively) and the items of the School functioning scale probably reflected too diverse aspects of school functioning to yield a reliable scale.

Children judged their quality of life in a different way as parents do as can be concluded

from the generally low correlations between the QLQC scores of the children and the parents. Children probably value several aspects of their life differently from their parents. Therefore, it is important to include the children's self-reports in HRQOL research. Although parents are important informants on the HRQOL of their children, conclusions based solely on parents reports may be incomplete. This conclusion is supported by several reviews on HRQOL in pediatric populations (Eiser, Havermans, Craft, & Kernaham, 1995; Mulhern et al., 1989; Rosenbaum & Saigal, 1995) and by a recent study on the proxy-problem in HRQOL research by Theunissen et al. (1998). Therefore it is regrettable that the number of HRQOL instruments including children's self-reports is still limited to the Munich Quality of Life Questionnaire (Bullinger, Von Mackensen, & Kirchberger, 1994) and the Child Quality of Life Questionnaire (Graham, Stevenson, & Flynn, 1997). The QLQC may in the future contribute to filling this gap.

There were several indications of adequate discriminative, convergent, and divergent validity of the QLQC. In contrast to what was expected, there were few differences between the study group and the comparison group. This aspect will be discussed further in the next section. The QLQC appeared to discriminate well between children with high levels of emotional and behavioral problems and children without. Additionally, there were moderate to high correlations between scales of the QLQC and well-validated instruments measuring comparable aspects of psychosocial functioning, and the Cognitive Functioning scale of the QLQC proved to be sensitive for the lower level of cognitive functioning of the study group compared to normative data.

Although it can be concluded that the QLQC has some adequate psychometric properties, several adjustments to the QLQC will have to be made to enhance reliability and validity. Items will have to be added to the physical functioning domain, especially because functional status is not well enough covered. It will also be necessary to improve the family functioning and school functioning scales within the social functioning domain.

9.3.2. The HRQOL of children with CAA compared to the comparison group

Compared to the children in the comparison group, children in the study group had lower quality of life in the physical functioning domain as reported by both parents and by the children. In the psychological functioning domain, the cognitive functioning of the study group children was lower than of the comparison group children as reported by each informant. The study group children reported more anxious-depressed feelings, more problems in relation to siblings, and more problematic school functioning. It should be noted that the effect sizes of the differences in scores between study group and comparison group did not exceed 5 % of the explained variance, which can be considered as low. Although there are indications of reduced HRQOL, it can be concluded that the HRQOL of most children with CAA can be considered as good. It might be argued that the psychometric properties of the QLQC were too limited to detect real differences. However, because the QLQC detected real differences in cognitive functioning and

several aspects of psychosocial functioning, and because other well-validated instruments to assess psychosocial functioning neither detected differences between the study and comparison group, this argument seems of limited value.

Within the study group there were very few differences in HRQOL between subgroups. There were no differences in HRQOL between children with different diagnoses, nor was the HRQOL influenced by early disease indicators such as length of the first hospitalization or number of operations. Only children with more than one major congenital anomaly had lower scores on the broad physical functioning domain scale compared to children with only one major anomaly, which can be explained by the greater medical complexity of these multiple anomalies causing more serious physical complaints and necessitating longer medical treatment. The relation between different parameters related to the medical condition on the one hand and psychosocial adjustment on the other hand will be discussed in section 9.6..

9.4. The physical, cognitive, and psychosocial functioning of children with CAA

Because the selection procedures for the children in the comparison group, used in the quality of life part of the study, differed for the different communities and because there is no insight in the reasons for refusal in the comparison group, this group might not be fully representative for children in the general population. So, in the next part of the study, the scores of the children with CAA on the assessment instruments were compared to normative data.

9.4.1. Physical functioning

Twenty-six per cent of the children ($n = 29$) had moderate to serious physical problems. Two children had serious hearing impairments, but otherwise no children were seriously physically handicapped. Six children with hernia diaphragmatica or esophageal atresia had chronic bronchitis, which caused also problems with physical endurance in some cases. The most frequent physical problems were gastrointestinal problems related to the congenital anomaly. Four children with an esophageal atresia had moderate swallowing problems. Thirteen children with anorectal malformations and four children with Hirschsprung's disease had moderate to serious problems with fecal incontinence. These figures are comparable to figures reported in the literature (Diseth, Bjørnland, Nøvik, & Emblem, 1997; Diseth & Emblem, 1996; Ginn-Pease et al., 1991; Ludman, Spitz, & Kiely, 1994).

Considering the seriousness of their initial medical condition and the high burden caused by lengthy and intensive medical treatment procedures, it can be concluded that children with CAA were functioning physically relatively good. Very few children had major impairments. The fecal continence problems for children with anorectal malformations can be considered as the most troublesome, but did not cause major functional impairment for the children.

9.4.2. Cognitive functioning

The mean IQ of the children was 91.5 which is almost 10 points lower than the norm of 100. This lower cognitive functioning was also reflected in several aspects of school functioning. Seventeen per cent of the children participated in special education compared to 4 % in the general population (Pijl, 1997). Compared to children in the general population a relatively large proportion of the children had low school competence as reported by parents and teachers. Thirteen per cent of the children obtained scores in the deviant range on the School Competence scale of the CBCL compared to 4.5 % in the general population, and 23 % of the children scored in the deviant range on the Academic Performance scale of the TRF compared to 14.5 % in the general population (Verhulst, Van Der Ende, & Koot, 1996 & 1997).

Several studies reported lower cognitive functioning for children with congenital diaphragmatic hernia (Davenport, Rivlin, D'Souza, & Bianchi, 1992; Nobuhara, Lund, Mitchell, Kharasch, & Wilson, 1996), with esophageal atresia (Dera, Mies, & Martinus, 1980), and for a group with mixed diagnoses (Kato et al., 1993). Others did not find lower cognitive functioning for children with esophageal atresia (Lindahl, 1984), for children with anorectal malformations (Dera et al., 1980; Ginn-Pease et al., 1991), and for a group with mixed diagnoses (Ludman, Spitz, & Lansdown, 1993). No other studies reported specifically on school functioning, but Hassink, Rieu, Brugman, and Festen (1994) found that the educational level of adults with anorectal malformations was lower than in the general population. The conclusion seems justified that children with CAA compared to children in the general population are at risk for lower cognitive functioning, learning problems, and possibly lower educational levels.

9.4.3. Psychosocial functioning

According to the parents and teachers 20 % to 30 % of the children showed elevated levels of emotional and behavioral problems compared to children in the general population. The proportion of the scores in the borderline or clinical range were higher than in normative samples on almost all CBCL syndrome scales, on three of the eight TRF syndrome scales, and on all CBCL and TRF broad band scales and total problem scores. Scores in the borderline range or clinical range on the CBCL or TRF can be considered as a valid indicator of the presence of psychopathology (Achenbach, McConaughy, & Howell, 1987; Verhulst, Van Der Ende, & Koot, 1996). These high proportions of children with scores in the deviant range stand somewhat in contrast to the finding that the mean scores on the CBCL and TRF did not differ from normative scores. In the literature similar results are reported. Ginn-Pease et al. (1991) and Ludman et al., (1994) also found elevated proportions of deviant CBCL scores for children with anorectal malformations or abdominal wall defects. In both studies the mean CBCL scores were not reported. Diseth et al. (1996) found that a high proportion of children with anorectal malformations had a psychiatric disturbance (58 %), while the mean CBCL scores were in the

normal range. So, a relatively high proportion of children with CAA show parent-reported psychosocial maladjustment while at the same time a relatively large proportion of these children may be functioning better than average. Both effects probably compensate for each other and cause a mean score for the whole group in the normal range. An explanation for this mechanism might be that a congenital anomaly can be viewed as a risk factor and at the same time as a protective factor for psychosocial maladjustment. On the one hand, the large amount of problems with which a child and its family with a congenital anomaly are confronted, especially in the first years of life, may make them vulnerable to psychosocial problems. On the other hand these problems may enhance their personal strengths and coping mechanisms with the effect that in later years they are less vulnerable to psychosocial problems. Which mechanism is working in an individual child and family probably depends on several factors within the child and its parents. To clarify these mechanisms and identify risk and protective factors is a major task for follow-up studies of children with physical disorders (Sloper & Turner, 1993). Some aspects of this problem will be discussed in a later part of the discussion.

Compared to the general population, the study group children themselves reported no more problems in psychosocial functioning. On the contrary, they indicated a higher self-esteem and fewer depressive problems. Very few studies on the long-term psychosocial functioning included children's self reports. Ginn-Pease et al. (1991) reported that the self-esteem for children with abdominal wall defects or anorectal malformations were in the normal range and Diseth et al. (1996 & 1997) reported scores in the normal range for adolescents with Hirschsprung's disease or anorectal malformations on the self-report version of the CBCL. Only Ludman et al. (1994) found higher depression scores for children and adolescents with anorectal malformations. It is possible that adolescents are at greater risk than school-aged children for emotional problems and negative self-esteem (Diseth et al., 1996; Ginn-Pease et al., 1991). Because our study group consisted of 8 to 12-year old children, it will be necessary to follow them into adolescence to know what will be the course of their psychosocial development. But presently, it can be concluded that the children in our study group experienced no more emotional problems or lower self-esteem than children in the general population and appeared to cope well with having a congenital anomaly and its negative physical consequences.

9.5. Family functioning and parental psychosocial functioning

Overall family functioning, the quality of the marital relationship, and the psychosocial functioning of the parents were all in the normal range. Except one study of children with Hirschsprung's disease which gave comparable results to ours (Diseth et al., 1997) no other studies assessed the family functioning or parental psychosocial functioning of children with CAA. It can be concluded that parents cope well with the burden of having to care for a child with a CAA. It is important to note that family problems should not too easily be expected solely

because the parents are confronted with a child with a major congenital anomaly. Families appear to have positive coping mechanisms which help them adapt to the difficulties they encounter when a child in the family has a serious medical problem (Kazak, Segal-Andrews, & Johnson, 1995).

9.6. Determinants of the cognitive and psychosocial functioning

9.6.1. Influence of medical parameters on cognitive functioning

Intelligence nor school functioning differed between different diagnostic groups. Medical parameters such as length of the first hospitalization or number of operations were not related to IQ or school functioning. However, the presence of a major associated congenital anomaly was negatively related to IQ. Children with a major associated congenital anomaly had a mean IQ of 76.9 compared to an IQ of 94.4 for children with no or minor associated congenital anomalies. Also, children with major congenital anomalies had a 44 % participation in special education. No other studies reported on the influence of the presence of an associated congenital anomaly on the cognitive functioning of CAA. Other studies did find relations between other medical factors such as number of operations or surgical stress and cognitive functioning (Kato et al., 1993; Ludman et al., 1993), which were not replicated in our study. From these results it can be argued that there may be two mechanisms by which the cognitive functioning of children with CAA is negatively influenced. First, prenatal factors, which are related to the presence of congenital anomalies, and second, postnatal factors such as surgical stress. Future prospective studies will have to clarify this issue further.

9.6.2. Influence of medical parameters on psychosocial functioning

Psychosocial functioning of the children was to a large degree independent of medical factors. Psychosocial functioning was not related to diagnosis. Neither were other medical factors such as length of the first hospitalization, number of operations, or presence of associated congenital anomalies. Also, present physical functioning as reflected by the number and seriousness of parent-reported physical complaints was not related to most measures except that there was a low correlation with the CBCL total problem score and the children's depression score. Within the groups of children with Hirschsprung's disease or anorectal malformations it was expected that fecal incontinence would negatively influence psychosocial adjustment. However, we failed to find a relation between seriousness of fecal continence problems and parent-reported or self-reported psychosocial functioning.

An important conclusion of this study is that children with CAA are at risk for psychosocial maladjustment irrespective of diagnostic category. It has to be recognized that a

priori expectations of the influence of certain physical problems (such as fecal continence problems) on maladjustment were not fulfilled. Factors which are responsible for the increased risk of maladjustment appear to be equally distributed over the different diagnostic categories.

9.6.3. Influence of psychosocial stress on psychosocial functioning

In this study the number of major life events and disease-related stress as reported by the parents and the children were employed as measures of psychosocial stress. Disease-related stress was defined as psychosocial problems which the children encountered directly related to their congenital anomaly.

Disease-related stress was more or less equally distributed over the different diagnostic groups. This is an indication of the fact that different aspects of the different diseases cause psychosocial stress. In the interviews it appeared that some children had problems with their operation scar, others were ashamed of having a visible although sometimes minor congenital anomaly, for example an anomaly of the feet, while still others were bothered by a lack of physical endurance in sports caused by chronic bronchitis, and others were ashamed about their fecal incontinence. Hypotheses raised by literature reports (Ginn-Pease et al., 1991) as well as clinicians in our hospital, stating that Hirschsprung's disease and anorectal malformations would cause more psychosocial stress than other anomalies because of the fecal continence problems in these groups, were not supported by our results.

Number of major life-events had low correlations with parent-reported and self-reported psychosocial adjustment. By contrast, moderate to high correlations were found between parent-reported disease-related stress and problem behavior as reported by parents and teachers, and between self-reported disease-related stress and self-esteem. This negative relation between disease-related stress and parent-reported and teacher-reported problem behavior may partly explain the high proportion of children with scores in the deviant range on these parameters. Although in some studies the relation between fecal continence problems and psychosocial adjustment was investigated (Diseth et al., 1996 & 1997, Ginn-pease et al., 1991; Ludman et al., 1993), this is the first study in which disease-related stress was identified as a risk factor for psychosocial maladjustment in all major CAA. These results corroborate the conclusion that attention should be paid to the psychosocial influence of all CAA on psychosocial functioning.

9.6.4. Influence of family functioning on psychosocial functioning

Several aspects of family functioning such as overall family functioning, but also the quality of the marital relationship, and psychosocial functioning of the mothers were related to psychosocial adjustment of the children. But of these, overall family functioning had the strongest relation with measures of parent-reported problem behavior and measures of self-reported self-esteem and depression. The results of this study underscore the strong relation between the children's

psychosocial adjustment and family functioning which is found in pediatric as well as in other populations (Dadds, 1995; Kazak et al., 1995; Wallander & Varni, 1998).

Family functioning is a neglected area in follow-up research of children with CAA. Only one study investigated aspects of family functioning, i.e. parental warmth and chronic family difficulties, in relation to psychosocial adjustment in a population of adolescents with Hirschsprung's disease (Diseth et al., 1997). They reported that the children's global psychosocial functioning was strongly related to parental warmth. In a multiple regression, this effect obscured the relation between fecal incontinence and global psychosocial functioning which was also found. So, in that study as in ours, aspects of family functioning were more strongly related to outcome than disease parameters, or mediated the effect of disease parameters. However, it should be beared in mind that the relation between family functioning and psychosocial adjustment of children is reciprocal. This implicates that problems in family functioning may cause psychosocial maladjustment and vice versa. In any case, clinicians and researchers in this field should be more aware of the importance of the relation between family functioning and a child's psychosocial adjustment.

9.7. Implications for future research

A strength of the present study was that the study group was unselected, included a broad range of diagnoses, and that a broad range of standardized instruments was used to assess psychosocial functioning of the children and the families including mother, father, child, and teacher reports. The study sample can be considered as a representative sample of children with a major CAA in view of comparable survival rates reported in the literature and of the overall response rate of 83.7 % of the surviving children. A relative weakness of the study was the partially retrospective and cross-sectional nature of the study. In a retrospective study it is very difficult to obtain optimal data on many aspects of the early physical functioning of the children. This may obscure important relations between early physical functioning and later physical and psychosocial functioning. As a consequence important clues for early intervention may be missed. A longitudinal study on the psychosocial development of children with congenital anomalies in general and congenital abdominal anomalies in particular, would open possibilities to assess important aspects of the physical and psychosocial functioning of the children, of the parent-child interaction, and family functioning from early on and at different ages in later development. In this way more insight would be gained in the developmental pathways of children with congenital anomalies.

This study has shown that cognitive functioning is reduced in children with CAA and that cognitive functioning is negatively influenced by the presence of associated major congenital anomalies. This should be taken into account in future studies. Measures for cognitive functioning should be incorporated and relations between medical parameters and cognitive functioning

should be further investigated. Second, as there was no relation between diagnosis and psychosocial adjustment, while there were several indications that children with CAA are at increased risk for psychosocial maladjustment, future studies should be aimed at the whole group of CAA and not only to one or two anomalies. Disease-related stress was found in this study to be an important predictor of psychosocial maladjustment. In future studies, efforts should be aimed at coming to a better operationalization and assessment of disease-related stress of CAA in order to come to a better understanding of the causal mechanisms in the relation between maladjustment and CAA. Finally, as family functioning appeared to be a major determinant of psychosocial adjustment in CAA, this aspect should be incorporated in future follow-up studies. Neglect of this aspect will give a too limited view on the psychosocial adjustment of children with CAA.

Based on the results of this study, the following recommendations for future research on the outcome of congenital or acquired medical conditions in childhood can be given (see also Koot & Bouman, in press):

1. Design prospective and longitudinal studies for children with congenital anomalies.
2. In view of the relatively few specific aspects in the psychosocial adjustment of children with congenital as well as acquired medical conditions, studies incorporating a greater diversity of anomalies should be planned.
3. Design a system in which medical factors such as presence of multiple congenital anomalies, seriousness of the neonatal condition, and neonatal or later surgical stress can be assessed in a standardized way.
4. Assess the following aspects of a child functioning in a standardized way:
 - a. physical functioning including physical complaints as well as functional status
 - b. the specific psychosocial stressors of the medical condition(s) under study
 - c. cognitive functioning
 - d. psychosocial functioning with special attention to:
 - presence of emotional and behavioral problems
 - self-esteem
 - social competence and quality of peer-relations.
5. Incorporate the parents' as well as the children's self-reports in the study.
6. Include measures of family functioning in the study.
7. Enhance cooperation between departments of pediatrics, pediatric surgery, and pediatric neurology on the one hand and departments of pediatric psychology and child and adolescent psychiatry on the other hand, in order to stimulate joint research projects on the outcome of children with acquired or congenital physical disorders.

9.8. Implications for clinical practice

Many of the aforementioned recommendations for research are equally valuable for the clinician. Many of the children with a major congenital anomaly visit the hospital for follow-up visits. For some children it is necessary to make frequent visits because of medical problems, while others come only once a year. Our study shows that it is important to follow all children over a long period, because as a group they are at risk for cognitive or psychosocial problems. Surely it will be important to pay attention to the physical functioning of these children, which after all is the primary task of a pediatric surgeon. But it will also be important to probe the cognitive and psychosocial functioning of these children. The question: "How are you?", often is not enough. This does not mean that all these children should be evaluated by a psychologist or child psychiatrist. It is often sufficient if a doctor asks simple but specific questions about school, the relation with peers, and relation with parents and siblings. At greatest risk for psychosocial maladjustment appeared to be those children who had a high amount of disease-related stress and who came from the most problematic families. These aspects should be taken into account in clinical interviews with children with CAA and their families. Because we showed that children often have a different view on their functioning than parents, it must be emphasized that the children themselves should be encouraged to answer, although they may often be somewhat reluctant to do this in the presence of parents. As has been shown in this study, there should be an alertness for cognitive and learning problems as well as problems in psychosocial functioning. And lastly, there should be an alertness for psychosocial problems with all children with CAA.

If cognitive, learning, or psychosocial problems are present, further guidance and support for these children often will be necessary. A pediatric hospital should be able to offer these children and their parents adequate psychosocial support by psychologists, social workers, or child and adolescent psychiatrists. Also concerning clinical practice, cooperation between departments of pediatric psychology and child and adolescent psychiatry and the department of pediatric surgery will be needed to optimize the care for these children and their families.

9.9. Conclusion

This follow-up study of children with CAA showed that these children are at risk for cognitive, learning, and psychosocial problems. We found that children who experienced psychosocial problems which were directly related to the congenital anomaly and children from more problematic functioning families were at the greatest risk for psychosocial maladjustment. Long-term follow-up programs are needed to follow all children with a CAA in their development into adulthood, so that problems are detected in time and the proper guidance and support can be given.

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Appendix A

Kwaliteit van Leven Vragenlijst voor kinderen

Hierna staat een lijst met vragen die er over gaan hoe het met je gaat. Deze vragen gaan over je gezondheid, maar ook over hoe je je voelt en hoe het thuis en op school gaat. De bedoeling is dat jij deze vragen gaat beantwoorden. Als je de vragen invult denk dan steeds eraan hoe het nu met je gaat of hoe het de afgelopen 2 maanden met je is gegaan. Je kunt bij iedere vraag steeds kiezen uit drie mogelijke antwoorden:

0 = Zelden of nooit 1 = Soms 2 = Altijd of meestal

Lees iedere vraag goed door. Wil je een rondje geven om de 2 als het altijd of meestal zo met je gaat, om de 1 als het soms zo met je gaat en om de 0 als het zelden of nooit zo met je gaat.

Bedenk dat er geen goede of foute antwoorden zijn.

Heb je broer(s) of zus(sen)? 0 ja 0 neen

Als je geen broer(s) of zus(sen) hebt, kun je de vragen die daar over gaan overslaan.

0	1	2	1.	Ik voel me gezond.
0	1	2	2.	Ik kan sport of gymnastiek goed volhouden.
0	1	2	3.	Ik heb een goede eetlust.
0	1	2	4.	Ik heb hoofdpijn.
0	1	2	5.	Ik heb buikpijn.
0	1	2	6.	Ik heb rugpijn.
0	1	2	7.	Ik ben duizelig.
0	1	2	8.	Ik ben moe.
0	1	2	9.	Ik ben misselijk.
0	1	2	10.	Ik heb moeite met poepen.
0	1	2	11.	Ik heb last van diarree.
0	1	2	12.	Ik voel me fit.
0	1	2	13.	Ik heb last van benauwdheid.
0	1	2	14.	Ik heb last van huiduitslag.
0	1	2	15.	Ik heb pijn in armen of benen.
0	1	2	16.	Ik kan goed lopen.
0	1	2	17.	Ik kan goed zien.
0	1	2	18.	Ik kan goed horen.
0	1	2	19.	Ik kan netjes schrijven.
0	1	2	20.	Ik kan goed knutselen.
0	1	2	21.	Ik kan alle aan lichamelijke activiteiten die ik wil, meedoen.
0	1	2	22.	Ik moet geholpen worden bij wassen of aankleden.
0	1	2	23.	Ik ben goed in sporten of gymnastiek.
0	1	2	24.	Ik ben gezond genoeg om naar school te kunnen.
0	1	2	25.	Ik ben gezond genoeg om op een sport te kunnen.
0	1	2	26.	Ik ben gezond genoeg om een hobby te kunnen doen.

Let op:

De vragen 27 tot en met 32 hoef je alleen te beantwoorden als je regelmatig bij een dokter komt. Als je niet regelmatig bij een dokter komt mag je ze overslaan en doorgaan met vraag 33.

Wil je aangeven hoe vaak je de afgelopen twee maanden naar een dokter moest:

0 Meer dan één keer per week. 0 Eén keer per week. 0 Eén keer per maand.

- | | | | | |
|-------|---|---|-----|--|
| 0 | 1 | 2 | 27. | Ik vind het vervelend om naar de dokter te gaan. |
| 0 | 1 | 2 | 28. | Ik vind de behandeling door de dokter vervelend of pijnlijk. |
| 0 | 1 | 2 | 29. | Ik heb een hekel aan de medicijnen, die ik moet gebruiken.
(niet invullen, als je geen medicijnen gebruikt) |
| 0 | 1 | 2 | 30. | Ik heb een hekel aan andere dingen (bijvoorbeeld prikken, dieet, klyasma's), die thuis moeten gebeuren. (niet invullen, als dat niet op jou van toepassing is) |
| 0 | 1 | 2 | 31. | Ik word door de dokter goed geholpen. |
| 0 | 1 | 2 | 32. | Ik voel me beter door de behandeling van de dokter. |
| 0 | 1 | 2 | 33. | Ik vind de behandeling van de dokter belangrijk. |
| 0 | 1 | 2 | 34. | Omdat ik naar de dokter moet, moet ik van school weg blijven. |
| 0 | 1 | 2 | 35. | Door de behandeling van een dokter, mag ik niet aan een sport deelnemen. |
| 0 | 1 | 2 | 36. | Door de behandeling van een dokter mag ik thuis bepaalde dingen niet doen. |
| 0 | 1 | 2 | 37. | Door de behandeling van een dokter mag ik op school bepaalde dingen niet doen. |
| <hr/> | | | | |
| 0 | 1 | 2 | 38. | Ik ben tevreden over mijn leven. |
| 0 | 1 | 2 | 39. | Ik heb genoeg goede vrienden of vriendinnen. |
| 0 | 1 | 2 | 40. | Ik denk vriendelijk over anderen. |
| 0 | 1 | 2 | 41. | Ik ben zenuwachtig. |
| 0 | 1 | 2 | 42. | Ik vind, dat ik er goed uit zie. |
| 0 | 1 | 2 | 43. | Ik doe dingen zonder goed na te denken. |
| 0 | 1 | 2 | 44. | Ik zie ertegen op om naar school te gaan. |
| 0 | 1 | 2 | 45. | Ik kan goed praten met mijn ouder(s). |
| 0 | 1 | 2 | 46. | Ik ben bang. |
| 0 | 1 | 2 | 47. | Ik voel me prettig in het gezin. |
| 0 | 1 | 2 | 48. | Ik ben verlegen. |
| 0 | 1 | 2 | 49. | Ik heb onaardige gedachten over anderen. |
| 0 | 1 | 2 | 50. | Ik kan goed leren. |
| 0 | 1 | 2 | 51. | Ik heb ruzie met mijn broer(s) of zus(sen). |
| 0 | 1 | 2 | 52. | Ik verveel me. |
| 0 | 1 | 2 | 53. | Ik word snel kwaad. |
| 0 | 1 | 2 | 54. | Mijn geheugen is goed. |
| 0 | 1 | 2 | 55. | Ik werk hard op school. |
| 0 | 1 | 2 | 56. | Ik heb het gevoel, dat ik minder waard ben dan andere kinderen. |
| 0 | 1 | 2 | 57. | Ik heb het gevoel, dat de juf of meester mij helpt, als het nodig is. |
| 0 | 1 | 2 | 58. | Ik doe onaardig tegen anderen. |
| 0 | 1 | 2 | 59. | Als iets tegen zit, kan ik daar goed overheen komen. |
| 0 | 1 | 2 | 60. | Ik kan mijn aandacht goed bij schoolwerk houden. |
| 0 | 1 | 2 | 61. | Ik heb ruzie met andere kinderen (niet broers/zussen). |
| 0 | 1 | 2 | 62. | Ik heb het gevoel dat genoeg mensen van me houden. |
| 0 | 1 | 2 | 63. | Ik ben boos op mijn ouder(s). |
| 0 | 1 | 2 | 64. | Ik plaag andere kinderen. |
| 0 | 1 | 2 | 65. | In mijn vrije tijd kan ik me goed bezighouden. |
| 0 | 1 | 2 | 66. | Ik voel me verdrietig. |
| 0 | 1 | 2 | 67. | Ik kan goed opschieten met mijn broer(s) of zus(sen). |

- | | | | | |
|---|---|---|------|---|
| 0 | 1 | 2 | 68. | Ik word geplaagd. |
| 0 | 1 | 2 | 69. | Ik ben achter op school. |
| 0 | 1 | 2 | 70. | Ik ben zeker van mezelf. |
| 0 | 1 | 2 | 71. | Ik maak me zorgen om wat familieleden zou kunnen overkomen. |
| 0 | 1 | 2 | 72. | Een belofte kom ik zeker na. |
| 0 | 1 | 2 | 73. | Ik heb ruzie met mijn ouder(s). |
| 0 | 1 | 2 | 74. | Ik vind dat ik de meeste dingen goed doe. |
| 0 | 1 | 2 | 75. | In mijn vrije tijd heb ik genoeg te doen. |
| 0 | 1 | 2 | 76. | Ik ben goed in rekenen. |
| 0 | 1 | 2 | 77. | Ik vind de juf of meester aardig. |
| 0 | 1 | 2 | 78. | Ik heb vertrouwen in mezelf. |
| 0 | 1 | 2 | 79. | Ik kan goed vrienden maken. |
| 0 | 1 | 2 | 80. | Ik ben ongehoorzaam. |
| 0 | 1 | 2 | 81. | Ik heb te weinig vrienden of vriendinnen. |
| 0 | 1 | 2 | 82. | Ik vind mijn leven waardeloos. |
| 0 | 1 | 2 | 83. | Ik ben in de war. |
| 0 | 1 | 2 | 84. | Ik heb vertrouwen in mijn ouder(s). |
| 0 | 1 | 2 | 85. | Ik ben liever alleen. |
| 0 | 1 | 2 | 86. | Ik klik. |
| 0 | 1 | 2 | 87. | Het gaat goed op school. |
| 0 | 1 | 2 | 88. | Ik kan goed opschieten met andere kinderen. |
| 0 | 1 | 2 | 89. | Ik ben bang om alleen te zijn. |
| 0 | 1 | 2 | 90. | Ik ben goed in taal. |
| 0 | 1 | 2 | 91. | Ik word geplaagd door mijn broer(s) of zus(sen). |
| 0 | 1 | 2 | 92. | Ik gedraag me goed op school. |
| 0 | 1 | 2 | 93. | Ik vind mezelf dommer dan andere kinderen. |
| 0 | 1 | 2 | 94. | Ik ben bang om aan clubs, die ik wel leuk vind, mee te doen. |
| 0 | 1 | 2 | 95. | Andere kinderen willen graag met mij spelen. |
| 0 | 1 | 2 | 96. | Ik voel me lusteloos. |
| 0 | 1 | 2 | 97. | Ik ben snel afgeleid. |
| 0 | 1 | 2 | 98. | Ik kan goed praten of spelen met mijn broer(s) of zus(sen). |
| 0 | 1 | 2 | 99. | Ik voel me eenzaam. |
| 0 | 1 | 2 | 100. | Ik schep op. |
| 0 | 1 | 2 | 101. | De meeste kinderen vinden mij aardig. |
| 0 | 1 | 2 | 102. | Ik ben trots op de dingen die ik goed doe. |
| 0 | 1 | 2 | 103. | Ik voel me op mijn gemak. |
| 0 | 1 | 2 | 104. | Ik lieg. |
| 0 | 1 | 2 | 105. | Ik ben tevreden met mezelf. |
| 0 | 1 | 2 | 106. | Ik ga graag naar school. |
| 0 | 1 | 2 | 107. | Ik luister slecht naar mijn ouder(s). |
| 0 | 1 | 2 | 108. | Ik heb zorgen. |
| 0 | 1 | 2 | 109. | Ik ben op school ongehoorzaam. |
| 0 | 1 | 2 | 110. | Ik voel me vrolijk. |
| 0 | 1 | 2 | 111. | Ik kan goed uit mijn woorden komen. |
| 0 | 1 | 2 | 112. | Ik heb overal zin in. |
| 0 | 1 | 2 | 113. | Ik doe wel eens, alsof ik ziek ben, om niet naar school te hoeven gaan. |
| 0 | 1 | 2 | 114. | Mijn broer(s) of zus(sen) helpen mij als dat nodig is. |
| 0 | 1 | 2 | 115. | Ik heb plezier in mijn leven. |
| 0 | 1 | 2 | 116. | Ik kan goed met mijn ouder(s) opschieten. |
| 0 | 1 | 2 | 117. | Ik voel me gelukkig. |
| 0 | 1 | 2 | 118. | Ik kan goed voor mezelf opkomen. |

Summary

The aim of this study was to assess how 8- to 12-year old children who are born with a major congenital abdominal anomaly, are functioning physically, psychologically, and socially and to assess by which medical or environmental factors this is influenced.

The birth of a child with a congenital anomaly places the parents as well as the medical team before a great number of questions. On the one hand, the parents will have questions about the cause of the anomaly. Questions which will not be answered, in most cases. Further, many parents will have great concerns about the medical situation of their child who in most cases will be hospitalized in a pediatric surgery intensive care unit. Finally, parents will be concerned about the future of their child and the consequences of the anomaly and the long-lasting and intensive treatment on his/her development.

The medical team will also be confronted with several questions during the treatment of children with major congenital abdominal anomalies. Many children with major congenital anomalies can be treated, although the treatment may be intensive and prolonged. However, for some children with multiple major congenital anomalies the treatment may be so intensive, while there is great uncertainty about the outcome, that the medical team may be confronted with serious ethical dilemmas.

To answer the questions of parents as well as the medical team, the literature concerning congenital anomalies was reviewed. From this literature review it can be concluded that children with congenital anomalies are at risk for psychosocial problems and that psychosocial functioning is influenced by environmental factors, such as family functioning. However, the research concerning children with major congenital abdominal anomalies appeared to be very scarce.

These considerations have instigated the departments of pediatric surgery and child and adolescent psychiatry of the Sophia Children's Hospital to start a follow-up study of children with major congenital abdominal anomalies. The questions of this study were:

1. How are 8- to 12-year old children with a major congenital abdominal anomaly functioning physically, psychologically, and socially?
2. Which medical parameters have a major influence on the outcome of these children, especially concerning their psychosocial functioning?
3. Which other factors, especially environmental factors, are related to the future psychosocial functioning of these children?
4. Which children run the greatest risk for problems in psychosocial functioning and are, therefore, in the greatest need of psychosocial guidance or support?

In chapter 2, an extensive literature review concerning the quality of life of children with acquired as well as congenital abdominal disorders is given. In this chapter, quality of life is defined as the level of physical, psychological, and social functioning as reported by the parents

or the children. The disorders described in this chapter are recurrent abdominal pain, inflammatory bowel diseases (Crohn's disease and ulcerative colitis), and congenital abdominal anomalies. Children with recurrent abdominal pain have, by definition, more physical complaints than children in the general population and these children are also limited in their functioning, for example in their participation in school or sports, but this aspect appeared to be scarcely studied. Several studies found that children with recurrent abdominal pain have more emotional problems (depression and anxiety) than children in the general population. Inflammatory bowel diseases cause a large amount of abdominal as well as extra-intestinal complaints. In the past it has been postulated that inflammatory bowel diseases are symptoms of severe underlying psychopathology, the so-called psychosomatic hypothesis. Carefully designed research showed that this hypothesis could not be maintained. However, several studies have shown that children with inflammatory bowel diseases have more emotional problems than children in the general population, which are probably caused by the burden of having a chronic disease. From the few studies which investigated the relation between seriousness of the disease and level of psychosocial functioning it appeared that there was no such a relation. Family functioning appeared to be hardly investigated in inflammatory bowel diseases and no conclusions can be drawn.

Congenital abdominal anomalies cause several physical, especially abdominal, complaints which seldom cause serious limitations. Lung problems are described in esophageal atresia and diaphragmatic hernia. Swallowing problems are described in esophageal atresia, and abdominal pain in abdominal wall defects and intestinal atresia. The most important physical problems in Hirschsprung's disease and anorectal malformations are constipation and fecal incontinence. Research of psychosocial functioning of children with congenital abdominal anomalies appeared to be very scarce. In congenital diaphragmatic hernia only the cognitive functioning has been investigated. In these studies it was shown that especially children who were treated with extra-corporeal membrane oxygenation (ECMO) are at risk for cognitive problems. Only one study has shown in a limited number of children with esophageal atresia that they have more emotional problems than children in the general population. Some studies, only concerning adults with esophageal atresia, found normal quality of life. Compared to the general population, a larger proportion of children with abdominal wall defects showed emotional and behavioral problems, while this was not found in children with intestinal atresia and Hirschsprung's disease. Most studies concerning psychosocial functioning of children with congenital abdominal anomalies are performed in children with anorectal malformations, but the results were contradictory. One study found a large number of psychiatric problems in a group of adolescents with anorectal malformations but no higher mean scores on standardized questionnaires for behavioral and emotional problems (Child Behavior Checklist, CBCL). Two other studies did find larger than expected proportions of children with anorectal malformations having emotional and behavioral problems. Based on this literature review, it can be concluded that there are indications that children with congenital abdominal anomalies have an increased risk for psychosocial problems, but that reliable studies are very scarce.

In chapter 3, the design of the study is discussed. The study group consisted of 139 unselected children, 87 boys and 52 girls, aged 8 to 12 years, with the six so-called 'index diagnoses' which together are the most frequent congenital abdominal anomalies. These anomalies are: esophageal atresia characterized by a malformation of the esophagus; diaphragmatic hernia characterized by a defect in the diaphragm leading to herniation of a the gut in the pleural cavity; abdominal wall defects characterized by herniation of the gut through the abdominal wall; intestinal atresia characterized by an occlusion of the intestines; Hirschsprung's disease characterized by an incomplete innervation of a part of the small or large intestines causing a functional stenosis; and anorectal malformations characterized by the anal canal ending in a blind loop. The instruments consisted of a physical examination and an abbreviated intelligence test of the children, and a psychosocial interview of the children and the parents concerning general psychosocial functioning and psychosocial stress directly related to the anomaly. In addition, several standardized questionnaires were completed including parent-reported and teacher-reported emotional and behavioral problems, self-reported depression and self-esteem, and parent-reported family functioning, marital relationship, and parental psychosocial functioning.

A part of the study concerned the assessment of the quality of life of the children. This part is described in chapter 4. Quality of life (QoL) is, in accord with a large part of the QoL research in pediatric populations, defined as a multidimensional construct comprising the physical, psychological, and social functioning domains. In this study it was thought to be important to obtain the parents' as well as the children's judgment on the children's QoL. A questionnaire was developed consisting of 118 questions to be answered on a three-point scale of which a parent and child version was made. The questionnaire, the Quality of Life Questionnaire for Children (QLQC) was completed by the children and the parents of the study group and by the parents and children of a comparison group consisting of 136 children matched with the study group for urbanization, age, and gender.

The results showed that children from the age of 8 and older were well able to complete the QLQC. Within the three domains, three to six scales could be constructed. The reliability, as measured with the internal consistency and the test-retest correlations, appeared to be satisfactory for the three overall domains scales and for most subscales especially in the psychological functioning domain. The reliability of some of the scales in the physical and social functioning domains were insufficient and should be improved in a next version. There appeared to be generally low correlations between the scores of the children and the parents, while the correlations between the mothers and the fathers were much higher. This can be regarded as a confirmation of the necessity to develop a self-report QoL instrument for children.

As measured with the QLQC, the study group showed poorer functioning in the physical domain and on the cognitive functioning scale of the psychological functioning domain. In other aspects there were only few differences between the study group and the comparison group. The QLQC discriminated well between children with high versus children with low CBCL problem

scores. Correlations between several scales of the QLQC and scales of other instruments measuring a comparable aspect of functioning were moderate to high, while correlations between non-conversing scales were low. The QLQC can be considered as a QoL instrument with good possibilities of which several aspects can be improved in future research.

In chapter 5, the outcome of children with a congenital diaphragmatic hernia are described. In this group were 11 children, 6 boys and 5 girls. About half of the children had, mostly minor, physical complaints. The mean IQ of the children was 85 which is 15 points lower than the norm of 100. It is indicated in the literature, that especially children with congenital diaphragmatic hernia who are treated with ECMO are at risk for cognitive problems. The results of this study showed that this may be the case for all children with a congenital diaphragmatic hernia. Significantly more children than expected (30 %) obtained scores in the borderline or clinical range of the CBCL, which is a clear indication for having emotional or behavioral problems. Two children had moderate psychosocial problems as indicated during the psychosocial interviews with the children and the parents.

The results of the children with esophageal atresia, 20 boys and 16 girls, are described in chapter 6. In children with esophageal atresia a high-risk group can be discerned who have a second major congenital anomaly and who have been dependent on artificial ventilation as a neonate and a low-risk group in which this was not the case. Eight children fell in the high-risk group. In esophageal atresia, the physical functioning is classified according to the so-called 'Desjardins' classification. According to this classification, the outcome of 16 children was excellent, of 9 children it was good, having only minor feeding or respiratory problems, and of 4 children fair, having moderate feeding or respiratory problems. These results are comparable to the results of other follow-up studies of children with esophageal atresia. There was no difference between the children in the high-risk group versus the other children. The IQ of the children was almost 10 points lower than the norm of 100 and 8 children (22 %) required special education which is five times as frequent as in the general population (4%). Children in the high-risk group had a mean IQ of 79.4, while the IQ of the children in the low-risk group children was 14 points higher (93.5). Twice as many children as in the general population obtained high scores for emotional and behavioral problems on the CBCL and the teacher's report form (TRF). High scores are defined as scores in the borderline or clinical range on the CBCL or TRF, which is a clear indication for having emotional or behavioral problems. The children themselves did not report more depressive problems or lower self-esteem than children in the general population, but children with lower intelligence reported more depressive problems. The level of family functioning and psychosocial stress were comparable to the general population. Children from poorer functioning families showed more emotional and behavioral problems.

In chapter 7, the results are described of the children with Hirschsprung's disease or anorectal malformations. The most important physical problems in these group were problems with fecal incontinence. It appeared that children with anorectal malformations had more problems with fecal incontinence than children with Hirschsprung's disease. Especially children

with associated urogenital malformations had serious continence problems. Also children with Hirschsprung's disease and anorectal malformations had lower mean IQ's and more children than expected participated in special education. Twice as many children as in the general population had CBCL and TRF scores in the borderline or clinical range. The children did not report higher depression scores or lower self-esteem. There appeared to be no differences in psychosocial functioning between children with Hirschsprung's disease or anorectal malformations, and neither between children with versus children without fecal incontinence. These results were against the expectations, because it was hypothesized that fecal incontinence would be such a burden for the children that they would show more psychosocial problems.

In chapter 8 the results of the entire group are described. The results of comparisons with normative data on psychosocial functioning of the children, family functioning, and parental functioning are given. Further, the association of the psychosocial functioning of the children with medical and environmental factors was studied. To this end, the 'disability-stress-coping model' of Wallander and Varni was used. In this model, the psychosocial functioning of children is brought in relation to risk factors such as aspects of the disease and psychosocial stress on the one hand, and to resistance factors such as intrapersonal factors and social-ecological factors such as parental or family functioning. Although 27 children had moderate physical problems, the physical functioning of the whole group can be described as good. With the exception of two children with serious hearing problems, no children had serious physical limitations. The mean IQ of the children was 8.5 points lower than the norm of 100 and four times as many children as in the general population participated in special education (17%). Thirty per cent of the children had scores in the borderline or clinical range on the CBCL and TRF. Depression and self-esteem scores were in the normal range. Family functioning, marital relationship, parental psychosocial functioning, and general psychosocial stress were all normal. It appeared that disease-related factors such as diagnosis or seriousness of physical complaints was only in a limited way related to psychosocial functioning of the children. However, it appeared that children with multiple major congenital anomalies had a much lower level of intelligence (mean IQ = 76.9) than children without (mean IQ = 94.4). The psychosocial functioning of the children appeared to be related to disease-related stress as reported by parents as well as children and to the level of family functioning. These factors explained together 17.1% to 35.1% of the variance in the children's psychosocial functioning. The level of disease-related stress was independent of the nature of the anomaly. The Wallander-Varni model appeared to be an important tool to investigate relations between outcome and determining factors and could partly be confirmed in the present study.

As discussed in chapter 9, the results of the present study lead to some important conclusions concerning the psychosocial functioning of children with congenital abdominal anomalies. Children with a congenital abdominal anomaly appeared to be at increased risk for psychosocial problems. These problems were independent of the nature of the anomaly, but more related to disease-related stress and family functioning. All congenital abdominal anomalies appear to cause specific psychosocial stress which may negatively influence psychosocial

functioning. The nature of the psychosocial stress differs depending on the anomaly. So, in some anomalies the stress consists of continence problems, in others of the large operation scar, while some other children are bothered by limitations in physical endurance caused by respiratory problems. In future follow-up studies these results will have to be taken in account and it will be important to investigate a broad sample including all major congenital abdominal anomalies and not just one or two.

From this study follow some recommendations for further research. Future research should be more of a prospective and longitudinal nature, the functioning of the children should be assessed in a more standardized way, parents and children should be taken as informants, and a broader range of disorders could be investigated in one study. Cooperation between departments of pediatric surgery, pediatrics, medical psychology, and child and adolescent psychiatry should be enhanced. For clinical practice, it seems important to acknowledge that the whole group of children with congenital abdominal anomalies may show psychosocial problems and that attention for all these children is needed. Special interest should be given to high-risk children, who are children with higher amounts of disease-related stress and coming from worse functioning families and who could be identified by direct questioning of the children and parents about these aspects. Follow-up studies into adulthood are needed to enlarge the insight in the development of these children.

Samenvatting

Het doel van het in dit proefschrift beschreven onderzoek was om vast te stellen hoe 8- tot 12-jarige kinderen die geboren zijn met een ernstige aangeboren darmafwijking, functioneren in lichamelijk, geestelijk en sociaal opzicht en om te bepalen door welke medische of omgevingsfactoren dit functioneren beïnvloed wordt.

De geboorte van een kind met een aangeboren afwijking stelt zowel ouders als het medisch team voor een groot aantal vragen. Ouders zullen enerzijds vragen hebben over de oorzaak van de aandoening. Vragen waarover zij in veel gevallen in het onzekere zullen blijven. Veel ouders zullen verder grote zorgen hebben over de medische toestand van hun pasgeboren kind dat in veel gevallen opgenomen moet worden in een kinderchirurgische intensive care unit. Tenslotte zullen zij zich bezorgd maken over de toekomst van hun kind en de gevolgen die de aandoening en de langdurige en intensieve behandeling kunnen hebben gehad op zijn/haar ontwikkeling.

Ook het medisch team komt voor diverse vragen te staan bij de behandeling van kinderen met ernstige aangeboren darmafwijkingen. Veel kinderen met ernstige aangeboren afwijkingen kunnen, weliswaar met intensieve en langdurige behandeling, goed behandeld worden. Echter voor sommige kinderen met meerdere aangeboren afwijkingen kan de behandeling zo ingrijpend zijn terwijl er grote onzekerheid is over de uiteindelijke uitkomst, dat het medische team voor belangrijke ethische dilemma's komt te staan.

Om antwoord te kunnen geven op de vragen die er rezen zowel van de kant van ouders als van de kant van de behandelend artsen ten aanzien van kinderen met aangeboren darmafwijkingen werd de bestaande literatuur onderzocht. Uit dit literatuuronderzoek kwam naar voren dat kinderen met aangeboren afwijkingen een groter risico lopen op psychosociale problemen en dat omgevingsfactoren zoals gezinsfunctioneren een belangrijke invloed daarop uitoefenen. Tegelijk werd duidelijk dat er maar in zeer beperkte mate onderzoek is verricht naar het latere psychosociale functioneren van kinderen met aangeboren darmafwijkingen.

Deze bevindingen hebben ertoe geleid dat de afdelingen kinderheekunde en kinder- en jeugdpsychiatrie van het SKZ een vervolgonderzoek bij kinderen met een aangeboren darmafwijking zijn gestart. De vraagstellingen van het onderzoek waren:

1. Hoe functioneren 8- tot 12-jarige kinderen met een ernstige aangeboren darmafwijking in lichamelijk, psychologisch en sociaal opzicht?
2. Welke medische parameters hebben een belangrijke invloed op de uitkomst, vooral ten aanzien van het psychosociale functioneren van deze kinderen?
3. Welke andere factoren, vooral omgevingsfactoren, zijn gerelateerd aan het toekomstig psychosociaal functioneren van deze kinderen?
4. Welke kinderen lopen het grootste risico op problemen in het psychosociaal functioneren en hebben als zodanig de grootste behoefte aan psychosociale begeleiding of steun?

In hoofdstuk 2 wordt een uitgebreid literatuuroverzicht gegeven met betrekking tot de kwaliteit van leven van kinderen met zowel verworven als aangeboren darmaandoeningen. Kwaliteit van leven wordt in dit hoofdstuk gedefinieerd als het niveau van functioneren op lichamelijk, psychisch en sociaal gebied zoals dat wordt beoordeeld door de ouders of de kinderen zelf. De aandoeningen die in dit hoofdstuk beschreven worden zijn recidiverende buikpijn, inflammatoire darmaandoeningen (ziekte van Crohn en colitis ulcerosa) en de aangeboren darmafwijkingen. Bij kinderen met recidiverende buikpijn zijn er, per definitie, meer lichamelijke klachten dan bij kinderen in de algemene bevolking en deze kinderen zijn ook meer beperkt in hun functioneren, bijvoorbeeld ten aanzien van deelname aan school of sportactiviteiten, maar dit aspect blijkt toch maar in beperkte mate goed onderzocht. In diverse onderzoeken is gevonden dat kinderen met recidiverende buikpijn meer emotionele problemen (depressie en angst) hebben dan kinderen in de algemene bevolking. De inflammatoire darmaandoeningen gaan gepaard met een grote hoeveelheid lichamelijke klachten, zowel buikklachten als andersoortige klachten. In het verleden is gepostuleerd dat deze aandoeningen uitingen zijn van een onderliggend psychiatrisch lijden, de zogenaamde psychosomatische hypothese. Uit zorgvuldig opgezet onderzoek bleek dat deze hypothese niet gehandhaafd kon worden. Diverse onderzoeken hebben echter wel aangetoond dat kinderen met deze aandoeningen vaker dan kinderen in de algemene bevolking emotionele problemen vertonen, welke waarschijnlijk vooral veroorzaakt worden door de belasting van het hebben van een chronische ziekte. Uit de weinige onderzoeken die onderzocht hebben of er een relatie was tussen ernst van de aandoening en psychosociaal functioneren kwam naar voren dat deze relatie er niet was. Gezinsfunctioneren bij inflammatoire darmaandoeningen blijkt nauwelijks goed onderzocht te zijn en conclusies hierover kunnen dus niet goed getrokken worden.

Aangeboren darmafwijkingen veroorzaken diverse klachten, vooral buikklachten, welke zelden ernstige beperking in het functioneren veroorzaken. Longproblemen worden beschreven bij hernia diafragmatica en oesofagusatresie. Slikproblemen komen regelmatig voor bij oesofagusatresie en buikpijnklachten bij buikwanddefecten en dunne darm atresie. De belangrijkste lichamelijke problemen bij de ziekte van Hirschsprung en anusatresie zijn obstipatie en onzindelijkheid voor ontlasting. Systematisch onderzoek naar het psychisch en sociaal functioneren van kinderen met aangeboren darmafwijkingen blijkt zeer beperkt. Bij hernia diafragmatica is eigenlijk alleen naar het cognitieve functioneren gekeken waarbij naar voren is gekomen dat vooral kinderen die behandeld zijn met extra corporele membraan oxygenatie (ECMO) risico lopen op een verlaagd IQ. Slechts één studie heeft bij een beperkt aantal kinderen met oesofagusatresie ($n = 10$) gevonden dat zij meer emotionele problemen vertoonden. Enkele studies, die alleen volwassenen met oesofagusatresie betroffen, vonden een normale kwaliteit van leven. In vergelijking met de algemene bevolking werd een groter aantal kinderen met emotionele en gedragsproblemen gevonden in een vervolgonderzoek van kinderen met buikwanddefecten, maar niet in onderzoeken naar dunne darm atresie en de ziekte van Hirschsprung. De meeste onderzoeken zijn gedaan naar het psychosociaal functioneren van

kinderen met anusatresie, maar deze vonden tegenstrijdige resultaten. Eén studie vond een groot aantal psychiatrische problemen in een groep adolescenten met een anusatresie, maar geen hogere gemiddelde scores op een gestandaardiseerde vragenlijst voor emotionele en gedragsproblemen bij kinderen (Child Behavior Checklist, CBCL). Twee andere studies vonden wel dat een groter dan verwacht gedeelte van deze kinderen emotionele en gedragsproblemen vertoonde. Op basis van dit literatuur overzicht kon geconcludeerd worden dat er weliswaar aanwijzingen zijn dat kinderen met aangeboren darmafwijkingen een hogere kans hebben op psychosociale problemen, maar dat betrouwbaar onderzoek ter zake zeer schaars is.

In hoofdstuk 3 wordt de opzet van het onderzoek besproken. De onderzoeksgroep bestond uit een groep van 139 niet geselecteerde kinderen, 87 jongens en 52 meisjes, in de leeftijd van 8 tot 12 jaar met de zes zogenaamde indexdiagnoses welke samen de belangrijkste ernstige aangeboren darmafwijkingen vormen. Deze afwijkingen zijn: oesofagusatresie, waarbij de slokdarm niet goed is aangelegd; hernia diafragmatica, waarbij een groot deel van de buikinhoud door een gat in het middenrif in de borstholte is terechtgekomen; buikwanddefecten, waarbij een gedeelte van de buikinhoud door de buikwand heen naar buiten uitpuilt; dunne darmatresie, waarbij de doorgang door de dunne darm is afgesloten; de ziekte van Hirschsprung, waarbij een gedeelte van de darm onvoldoende geïnnerveerd is waardoor er een functionele vernauwing ontstaat; en anusatresie, waarbij de anus blind eindigt en er geen uitgang naar buiten is. De meetinstrumenten worden beschreven. Naast een lichamelijk onderzoek en een verkort intelligentieonderzoek van de kinderen, een psychosociaal interview van de kinderen en de ouders met betrekking tot algemeen psychosociaal functioneren en ervaren psychosociale stress in relatie tot de aandoening, werden diverse gestandaardiseerde instrumenten met betrekking tot het psychosociaal functioneren van de kinderen en de ouders afgenomen.

Een deel van het onderzoek stond in het teken van het bepalen van de kwaliteit van leven van de kinderen. Dit wordt beschreven in hoofdstuk 4. Kwaliteit van leven (KvL) is, in overeenstemming met een groot deel van het KvL onderzoek bij kinderen met lichamelijke aandoeningen, in dit onderzoek gedefinieerd als een multidimensioneel construct waarin het functioneren op lichamelijk, psychologisch en sociaal gebied vervat is. Het werd binnen dit onderzoek van belang geacht om de beoordeling van de KvL van de kinderen zowel door de ouders als door hen zelf te laten geschieden. Er werd dus een vragenlijst ontwikkeld bestaande uit 118 vragen die beantwoord konden worden op een schaal van 0 tot 2, en waarvan zowel een ouderversie als een kinderversie werd gemaakt. De vragenlijst, de Kwaliteit van Leven Vragenlijst voor Kinderen (KLVK), werd voorgelegd aan de kinderen en de ouders van de onderzoeksgroep en aan een vergelijkingsgroep van 136 kinderen die overeen kwamen met de onderzoeksgroep met betrekking tot urbanisatiegraad, leeftijd en geslacht.

Uit de resultaten bleek dat kinderen vanaf 8 jaar de KLVK goed kunnen invullen. Er konden binnen de drie domeinen 3 tot 6 schalen geconstrueerd worden. De betrouwbaarheid van de KLVK, zoals bepaald met de mate van interne consistentie en de test-hertest betrouwbaarheid, bleek bevredigend voor de 3 overall domein schalen en voor de meeste subschalen vooral in het

psychologische domein. De betrouwbaarheid van enkele schalen in het lichamelijke en sociale domein was onvoldoende en deze schalen moeten in een volgende versie uitgebreid en verbeterd worden. Er bleken over het algemeen lage correlaties tussen de scores van de kinderen en die van de ouders terwijl de correlaties tussen de ouders onderling beduidend hoger waren. Dit kan worden beschouwd als een bevestiging van de noodzaak om een door kinderen zelf in te vullen KvL instrument te ontwikkelen. Gemeten met de KLVK bleek de onderzoeksgroep vooral slechter te functioneren in het lichamelijke domein en op de cognitieve schaal in het psychologische domein. Overigens waren er geen grote verschillen tussen beide groepen gemeten met de KLVK. De KLVK bleek wel goed onderscheid te maken tussen kinderen met een hoge CBCL probleemscore ten opzichte van kinderen met een lage score. Correlaties tussen diverse schalen van de KLVK en schalen van gestandaardiseerde instrumenten die een vergelijkbaar aspect van functioneren meten bleken matig tot hoog, terwijl correlaties tussen niet overeenkomende schalen laag bleken te zijn. De KLVK kan beschouwd worden als een KvL instrument met goede mogelijkheden waarvan echter diverse aspecten verbeterd kunnen worden en die in een vervolgonderzoek opgenomen kunnen worden.

In hoofdstuk 5 wordt de uitkomst beschreven voor de kinderen met een hernia diafragmatica. Dit waren 11 kinderen, 6 jongens en 5 meisjes. Ongeveer de helft van de kinderen had meestal lichte lichamelijke klachten. Het gemiddelde IQ van deze kinderen was 85, wat 15 punten lager is dan het normgemiddelde van 100. In de literatuur wordt aangegeven dat vooral kinderen met een hernia diafragmatica die behandeld zijn met ECMO risico lopen op een cognitieve achterstand. De uitkomst van dit onderzoek laat zien dat dat voor alle kinderen met een hernia diafragmatica kan gelden. Beduidend meer kinderen dan verwacht (30 %) behaalden scores in het borderline of klinische gebied op de CBCL, wat een duidelijke indicatie is voor het hebben van emotionele of gedragsproblemen. Twee kinderen hadden matige psychosociale problemen zoals aangegeven door henzelf en de ouders gedurende de interviews.

De resultaten van de kinderen met een oesofagusatresie, 20 jongens en 16 meisjes, worden beschreven in hoofdstuk 6. Bij een oesofagusatresie kan onderscheid gemaakt worden tussen een hoog-risico groep met kinderen die een ernstige tweede aangeboren afwijking hebben en die als pasgeborene beademd zijn en een laag-risico groep waarvoor dat niet geldt. Acht kinderen in deze groep vielen in de hoog-risico groep. Het lichamenlijk functioneren wordt bij een oesofagusatresie ingedeeld volgens de zogenaamde 'Desjardins' classificatie. Volgens deze classificatie was van 16 kinderen de uitkomst uitstekend, van 9 kinderen goed, dat wil zeggen alleen lichte voedings- of ademhalingsproblemen, en van 4 kinderen matig, dat wil zeggen matige voedings- of ademhalingsproblemen. Deze uitkomst komt overeen met andere follow-up onderzoeken van kinderen met een oesofagusatresie. Er was geen verschil in lichamenlijk functioneren tussen kinderen in de hoog-risico groep ten opzichte van de andere kinderen. Het IQ van de kinderen was bijna 10 punten lager dan de norm van 100 en acht kinderen (22 %) volgden speciaal onderwijs wat meer dan vijf maal zoveel is als in de algemene bevolking (4 %). Kinderen in de hoog-risico groep hadden een gemiddeld IQ van 79.4, terwijl het gemiddelde IQ van de kinderen

in de laag-risico groep 14 punten hoger was (93.5). Twee maal zoveel kinderen als in de algemene bevolking hadden scores in de borderline of klinische range op de CBCL en de leerkracht vragenlijst (TRF). De kinderen zelf rapporteerden niet meer depressieve klachten of een lager zelfbeeld dan kinderen in de algemene bevolking, maar kinderen met een lagere intelligentie hadden meer depressieve klachten. Het niveau van gezinsfunctioneren en de mate van psychosociale stress waren vergelijkbaar met gegevens uit de algemene bevolking. Kinderen uit slechter functionerende gezinnen bleken meer emotionele en gedragsproblemen te vertonen.

In hoofdstuk 7 worden de resultaten beschreven van de kinderen met de ziekte van Hirschsprung en anusatresie. De belangrijkste lichamelijke problemen bij deze groep waren problemen met de continentie voor ontlasting. Het bleek dat kinderen met een anusatresie meer continentieproblemen vertoonden dan de kinderen met de ziekte van Hirschsprung. Vooral bij de kinderen die ook aangeboren urogenitale afwijkingen hadden waren de continentieproblemen het ernstigst. Ook in deze groep bleek het IQ gemiddeld lager dan in de algemene bevolking en zaten meer kinderen dan verwacht in het speciaal onderwijs. Twee maal zoveel kinderen hadden CBCL en TRF scores in het borderline of klinische gebied. Er waren geen hogere depressiescores of een lager zelfbeeld in deze groep kinderen. Er bleken geen verschillen te zijn in psychosociaal functioneren tussen de kinderen met de ziekte van Hirschsprung of anusatresie en evenmin tussen de kinderen met of zonder continentieproblemen. Deze laatste uitkomst was tegen de verwachting, aangezien tevoren verondersteld was dat de incontinentie een zodanige belasting voor de kinderen zou vormen dat zij meer psychosociale problemen zouden vertonen.

In hoofdstuk 8 worden de resultaten van de gehele groep besproken. Het psychosociaal functioneren van de kinderen en het gezinsfunctioneren en het functioneren van de ouders werd vergeleken met dat van de algemene bevolking. Daarnaast werd het psychosociaal functioneren van de kinderen in verband gebracht met zowel medische als psychosociale achtergrondgegevens. Bij dit laatste werd gebruik gemaakt van het zogenaamde 'disability-stress-coping model' van Wallander en Vami. In dit model wordt het psychosociaal functioneren van kinderen enerzijds in verband gebracht met risicofactoren, zoals aspecten van de aandoening en psychosociale stress, en anderzijds met beschermende factoren samenhangend met intrapersoonlijke factoren en psychosociale achtergrondfactoren, zoals ouderlijk of gezinsfunctioneren.

Hoewel 27 kinderen matige lichamelijke problemen hadden, kan het lichamelijk functioneren over de gehele groep goed genoemd worden. Behoudens twee kinderen met gehoorsproblemen, hadden geen van de kinderen ernstige functionele beperkingen. Over de gehele groep bleek het IQ 8,5 punt lager dan de norm van 100 en was deelname in speciaal onderwijs 4 maal zo hoog als in de algemene bevolking (17 %). Dertig procent van de kinderen hadden CBCL en TRF scores in het borderline of klinische gebied. Depressie- en zelfbeeldscores waren niet hoger dan bij kinderen in de algemene bevolking. Gezinsfunctioneren, echtpaarrelatie van de ouders, ouderlijk psychosociaal functioneren, en algemene psychosociale stress waren alle in het normale bereik. Het bleek dat ziekte-gerelateerde factoren zoals diagnose of ernst van de lichamelijke klachten bij follow-up slechts in zeer beperkte mate gerelateerd waren aan het

psychosociaal functioneren van de kinderen. Wel bleken kinderen met multiple ernstige aangeboren afwijkingen een beduidend lager intelligentie (IQ = 76,9) te hebben dan kinderen met maar een ernstige aangeboren afwijking (IQ = 94,4). Psychosociaal functioneren van de kinderen bleek vooral gerelateerd te zijn aan door zowel ouders als kinderen zelf aangegeven psychosociale stress die direct in verband stond met de aangeboren afwijking, en aan het niveau van gezinsfunctioneren zoals dat door de ouders was gerapporteerd. Deze beide factoren verklaarden 17,1 % tot 35,1 % van de variantie in het psychosociaal functioneren van de kinderen. De mate van door de aandoening veroorzaakte stress bleek onafhankelijk te zijn van de aard van de aandoening. Het model van Wallander en Varni bleek een belangrijk hulpmiddel om relaties tussen uitkomst en bepalende factoren te onderzoeken en kon in het huidige onderzoek gedeeltelijk bevestigd worden.

De bevindingen van dit onderzoek leiden tot enkele belangrijke conclusies met betrekking tot het psychosociaal functioneren van kinderen met een aangeboren darmafwijking welke besproken worden in hoofdstuk 9. Het bleek zo te zijn dat kinderen met een ernstige aangeboren darmafwijking een verhoogd risico lopen op problemen op psychosociaal gebied. Deze problemen zijn echter onafhankelijk van de aard van de aandoening, maar hangen vooral samen met aan de aandoening gerelateerde stress en gezinsfunctioneren. Alle aangeboren darmafwijkingen blijken dus zodanige stress te veroorzaken dat het psychosociaal functioneren daardoor negatief beïnvloed wordt. Uiteraard is het zo dat de aard van de stress per aandoening verschilt, zodanig dat bij de ene aandoening het de continentieproblemen zijn die stress veroorzaken, maar bij de andere het grote litteken of beperkingen in het uithoudingsvermogen tengevolge van longproblemen. Bij toekomstig vervolgonderzoek zal hiermee rekening gehouden moeten worden en is het belangrijk om alle aangeboren darmafwijkingen daarin te betrekken en niet alleen een beperkt aantal diagnoses.

Uit dit onderzoek volgt een aantal aanbevelingen voor verder onderzoek die erop neerkomen dat vervolgonderzoek meer prospectief en longitudinaal van aard zou moeten zijn, dat op een gestandaardiseerde manier naar diverse aspecten van het functioneren van de kinderen moet worden gekeken, dat het oordeel van zowel kinderen als ouders meegenomen moet worden en dat een groter scala aan aandoeningen in één onderzoek betrokken zou kunnen worden. Nauwere samenwerking op dit gebied tussen afdelingen kinderheelkunde, kindergeneeskunde, medische psychologie en kinder- en jeugdpsychiatrie wordt bepleit. Voor de klinische praktijk lijkt het van groot belang om te onderkennen dat de gehele groep kinderen met aangeboren darmafwijkingen psychosociale problemen kunnen gaan vertonen en dat aandacht hiervoor bij follow-up van deze kinderen van belang is. Daarbij zouden kinderen met een verhoogd risico afkomstig uit meer problematische gezinnen en met een hogere ervaren aandoening gerelateerde stress, door gerichte vragen eerder geïdentificeerd kunnen worden. Vervolgonderzoek tot in de volwassenheid ook van de huidige groep is noodzakelijk om het inzicht in de ontwikkeling van deze kinderen te vergroten.

Résumé

L'objectif de cette étude était de déterminer le niveau de fonctionnement physique, psychique et social d'enfants, âgés de 8 à 12 ans, avec des anomalies congénitales abdominales majeures et de spécifier par quels facteurs médicaux ou sociaux ce fonctionnement est influencé.

La naissance d'un enfant avec une anomalie congénitale confronte les parents aussi bien que l'équipe médicale avec un grand nombre de questions. D'une part, les parents auront des questions sur la cause de l'anomalie. Des questions qui ne seront probablement pas résolues. Beaucoup de parents auront des questions sur l'état médical de leur enfant qui est souvent hospitalisé dans un centre de soins intensifs pour chirurgie d'enfant. Enfin, les parents se feront des soucis sur l'avenir de leur enfant et sur les conséquences que l'anomalie et le traitement peuvent avoir sur le développement de leur enfant.

L'équipe médicale aussi est confrontée avec plusieurs questions concernant le traitement d'enfants avec des anomalies congénitales abdominales majeures. Il y a souvent de bonnes possibilités de traiter les enfants avec des anomalies congénitales abdominales majeures, mais parfois le traitement nécessaire peut être si drastique et le résultat si incertain que l'équipe médicale se voit posé devant des dilemmes éthiques.

Pour répondre aux questions des parents ainsi que celles des médecins, nous avons étudié la littérature existante concernant des enfants avec des anomalies congénitales. De cette recherche de la littérature ressort que des enfants avec des anomalies congénitales ont un risque élevé de problèmes psychosociaux et que le fonctionnement familial est un déterminant important de ces problèmes. En même temps, il est devenu clair que la recherche sur le fonctionnement psychosocial des enfants avec des anomalies congénitales abdominales est rare.

Ces données ont stimulé les départements de chirurgie d'enfant et de psychiatrie d'enfant pour commencer une recherche sur des enfants ayant des anomalies congénitales abdominales majeures. Les questions que cette recherche se pose sont les suivantes :

1. Comment est le fonctionnement physique, psychique et social d'enfants âgés de 8 à 12 ans avec des anomalies congénitales abdominales majeures?
2. Quels facteurs médicaux ont une influence importante sur ce fonctionnement?
3. Quels autres facteurs non-médicaux se relient au fonctionnement psychosocial futur de ces enfants?
4. Quels enfants ont le plus grand risque de problèmes psychosociaux et ont le plus besoin de support psychosocial?

En chapitre 2 la littérature concernant la qualité de vie d'enfants avec des maladies abdominales acquises ou congénitales est rapportée. 'La qualité de vie' est définie par le niveau

de fonctionnement dans le domaine physique, psychique et social, jugé par les parents ou les enfants eux-mêmes. Les maladies, qui sont décrites dans ce chapitre, sont: douleurs abdominales récurrentes, les maladies inflammatoires du tube digestif (maladie de Crohn et colite ulcéraire) et les anomalies congénitales abdominales. Par définition, les enfants avec douleurs abdominales récurrentes ont des problèmes physiques plus fréquents que des enfants dans la population générale. Ces enfants sont aussi plus limités dans leurs activités scolaires ou sportives, mais cet aspect n'est recherché que d'une manière limitée. Plusieurs recherches ont montré, que des enfants avec douleurs abdominales récurrentes ont plus de problèmes émotionnels (dépression et anxiété) que des enfants dans la population générale. Les maladies inflammatoires du tube digestif causent beaucoup de plaintes physiques, abdominales aussi bien que non-abdominales. Dans le passé, il est postulé que ces maladies sont l'expression d'une maladie psychiatrique, que l'on appelle l'hypothèse psychosomatique. Des recherches méticuleuses ont montré que cette hypothèse ne pouvait pas être défendue. Mais plusieurs recherches ont montré que les enfants souffrant de maladies inflammatoires du tube digestif ont plus de problèmes émotionnels que les enfants dans la population générale. Ces problèmes sont probablement causés par le poids d'une maladie chronique. Peu de recherches ont étudié la relation entre la gravité de ces maladies et le fonctionnement psychosocial. Ces recherches n'ont pas trouvé une relation entre les deux. Le fonctionnement familial a été très peu recherché et des conclusions à ce sujet ne peuvent donc pas être tirées.

Les anomalies congénitales abdominales causent des problèmes physiques divers. Tout d'abord, les problèmes abdominaux ne causent pas souvent de limitations sérieuses. Ensuite, des problèmes pulmonaires sont décrits en cas de la hernie diaphragmatique ou de l'atrésie de l'oesophage. Des problèmes de déglutition sont assez fréquents en cas de l'atrésie de l'oesophage et des douleurs abdominales en cas de malformations de la paroi abdominale ainsi que de l'atrésie intestinale. Les problèmes les plus graves en cas de la maladie de Hirschsprung et d'atrésie anorectale sont la constipation et l'incontinence fécale. Des recherches systématiques visant au fonctionnement psychique et social d'enfants ayant des anomalies congénitales abdominales sont très rares. Concernant la hernie diaphragmatique seulement le fonctionnement cognitif a été l'objet de quelques recherches. Dans ces recherches il est apparu que surtout les enfants avec une hernie diaphragmatique qui est traitée avec l'oxygénation extracorporelle membranaire courent le risque d'un quotient d'intelligence baissé. Une étude, faite auprès d'un nombre limité ($n = 10$) d'enfants ayant une atrésie de l'oesophage, a pu observer que ces enfants ont plus de troubles émotionnels. Quelques rares études qui concernaient seulement des adultes, ont trouvé une qualité de vie normale. Des études de suite ont donné comme résultat que, en comparaison avec la population générale, un plus grand nombre d'enfants avait des troubles du comportement et émotionnels en cas de malformations de la paroi abdominale mais pas dans le cas de l'atrésie intestinale ou de la maladie de Hirschsprung. La plupart des recherches avait porté sur le

fonctionnement psychosocial d'enfants ayant une atrésie anorectale, mais ces études ont donné des résultats contradictoires. Une seule étude a trouvé un grand nombre de problèmes psychiatriques dans un groupe d'adolescents avec une atrésie anorectale mais les scores obtenus par questionnaires standardisés portant sur les troubles du comportements ou émotionnels (Child Behavior Checklist, CBCL) furent peu élevés. Deux autres études par contre ont constaté qu'un nombre d'enfants dépassant l'expectative avait des troubles du comportement ou émotionnels. De ce résumé de la littérature on peut conclure que les enfants souffrant d'anomalies congénitales abdominales ont un plus grand risque de problèmes psychosociaux mais les recherches fiables concernant ce domaine sont très rares.

En chapitre 3, le dessein de l'étude est exposé. Les sujets de notre échantillon étaient: 139 enfants non-sélectionnés, 87 garçons et 52 filles, âgés de 8 à 12 ans avec les 6 diagnoses dites 'index': atrésie de l'oesophage, qui est caractérisée par une malformation de l'oesophage; hernie diaphragmatique congénitale, qui est caractérisée par un trou dans le diaphragme par lequel une grande partie de l'abdomen est 'herniée' dans le thorax; malformations de la paroi abdominale, qui sont caractérisées par une herniation du contenu de l'abdomen en dehors de la cavité abdominale; atrésie intestinale, qui est caractérisée par une occlusion du lumen des intestins; la maladie de Hirschsprung, qui est caractérisée par une déficience de l'innervation d'une partie du système intestinal causant une sténose fonctionnelle; et enfin malformation anorectale, qui est caractérisée par un anus clos. La méthodologie de l'étude comprenait: tout d'abord un examen physique, un test d'intelligence abrégé des enfants et un interview psychosocial des enfants et des parents concernant le fonctionnement psychosocial général et le stress psychosocial directement lié à l'anomalie. Ensuite un questionnaire standardisé concernant les troubles du comportement et émotionnels des enfants, rapportés par les parents et les instituteurs (CBCL et TRF), un questionnaire concernant les problèmes dépressifs et l'image de soi, rapportés par les enfants, et enfin un questionnaire concernant le fonctionnement de la famille, la relation entre les parents et le fonctionnement psychosocial des parents, rapportés par les parents.

La mise au point de la 'qualité de vie' était un des buts principaux de notre étude et est exposée en chapitre 4. En accord avec la majorité des recherches auprès d'enfants ayant des troubles somatiques, la qualité de vie est définie par le niveau de fonctionnement dans le domaine physique, psychique et social. Dans notre étude, il nous a semblé important d'obtenir le jugement aussi bien des parents que des enfants sur la qualité de vie des enfants de notre échantillon. Un questionnaire a été élaboré consistant de 118 questions. Les réponses sont cotées de 0 à 2. Une version pour les parents et une autre pour les enfants a été construite. Le questionnaire, dit le 'Questionnaire de Qualité de Vie pour Enfants' (QQVE), fut rempli d'une part par les enfants de l'échantillon d'étude et d'autre part par un groupe de comparaison de 136 enfants d'une population générale. Les enfants du groupe de comparaison concordaient avec les enfants du groupe de l'étude en ce qui concerne le degré d'urbanisation de leur domicile, leur âge et leur

sexe.

Des enfants dès l'âge de 8 ans étaient capables de remplir le questionnaire. Dans les 3 domaines, entre 3 et 6 échelles pouvaient être construites. La fidélité du QQVE, mesurée par le degré de consistance interne et la fidélité de test-retest, fut satisfaisante pour les échelles construites pour les 3 domaines de fonctionnement généraux et la fidélité fut également satisfaisante pour plusieurs échelles dans le domaine psychique et social. La fidélité de certaines échelles dans le domaine physique et social ne fut pas suffisante et ces échelles devront être remaniées dans une prochaine version. Les corrélations entre les scores des enfants et ceux des parents étaient basses tandis que les corrélations entre les scores des pères et ceux des mères étaient bien plus hautes. Ces résultats nous ont donné la confirmation de la nécessité d'élaborer un instrument pour mesurer la qualité de vie qui pourrait être rempli par les enfants eux-mêmes. Les résultats du QQVE ont montré que les enfants du groupe de l'étude fonctionnent moins bien que les enfants du groupe de comparaison notamment en ce qui concerne le domaine du fonctionnement physique et celui du développement cognitif. D'ailleurs, quand on mesure avec le QQVE, on ne trouve pas de grandes différences entre les deux groupes. Le QQVE différenciait bien entre les enfants obtenant des scores élevés sur le CBCL et les enfants obtenant des scores plus bas. Les corrélations entre plusieurs échelles du QQVE et les échelles d'autres instruments standardisés, qui mesurent un aspect du fonctionnement comparable, étaient modérées à hautes et les corrélations entre échelles non-correspondantes étaient basses. Le QQVE peut être considérée comme un instrument de qualité de vie ayant de bonnes possibilités. Cependant, certaines améliorations peuvent être introduites dans le QQVE, qui pourraient être incorporé dans une prochaine étude.

En chapitre 5, les résultats des enfants avec une hernie diaphragmatique congénitale, notamment 11 enfants, 6 garçons et 5 filles, sont décrits. Environ la moitié des enfants avait des problèmes physiques, plutôt légers. Le quotient d'intelligence moyen de ces enfants était de 85, ce qui est 15 points plus bas que la norme de 100. Dans la littérature, on constate que ce sont surtout les enfants avec une hernie diaphragmatique congénitale ayant été traités par oxygénation extracorporelle membraneuse qui courent le risque d'un retard cognitif. Les résultats de notre étude ont montré que tous les enfants avec une hernie diaphragmatique congénitale ont le risque d'un retard cognitif. Deux fois plus d'enfants avaient un niveau élevé de troubles du comportement ou émotionnels. Un 'niveau élevé' est défini ici par des scores dans la région 'borderline' ou 'clinique' sur le CBCL, ce qui est une indication que cet enfant a des troubles du comportement ou émotionnels. Deux enfants avaient des problèmes psychosociaux modérés rapportés par les parents et les enfants pendant les interviews.

Les résultats des enfants avec une atrésie de l'oesophage, 20 garçons et 16 filles, sont décrits en chapitre 6. En cas d'une atrésie de l'oesophage une distinction peut être faite entre un groupe à 'haut risque' comprenant des enfants ayant une deuxième anomalie congénitale

majeure et qui ont été dépendants de ventilation artificielle dans la période néonatale, et un autre groupe à 'bas risque' comprenant des enfants pour qui ces deux facteurs n'avaient pas été présents. Le fonctionnement physique en cas d'atrésie de l'oesophage est classifié selon la classification de Desjardins. Selon cette classification, 16 enfants fonctionnaient d'une manière excellent, 9 enfants fonctionnaient bien, ce qui veut dire qu'ils avaient des problèmes d'alimentation ou de respiration légers, et 4 enfants fonctionnaient d'une manière médiocre, ce qui veut dire qu'ils avaient des problèmes d'alimentation ou de respiration modérés. Ces résultats sont en accord avec d'autres études de suite d'enfants avec une atrésie de l'oesophage. Physiquement, il n'y avait pas de différences entre les enfants dans le groupe à 'haut risque' et le groupe à 'bas risque'. Le quotient d'intelligence moyen de ces enfants était de 10 points plus bas que la norme de 100 et 8 enfants (22 %) participaient à l'éducation spéciale, ce qui est plus de 5 fois plus que dans la population générale. Les enfants du groupe à 'haut risque' avaient un quotient d'intelligence moyen de 79,4, tandis que le quotient d'intelligence des enfants du groupe à 'bas risque' était de 14 points plus haut (93,5). Deux fois plus d'enfants avec une atrésie de l'oesophage (30 %) que des enfants de la population générale (15 %) avaient des scores élevés pour les troubles du comportement et émotionnels sur le CBCL et sur le questionnaire correspondant avec le CBCL pour les instituteurs (TRF). Les enfants ne rapportaient pas plus de problèmes dépressifs ni un image de soi plus bas que les enfants dans la population générale. Par contre, les enfants avec un quotient d'intelligence plus bas avaient plus de problèmes dépressifs. Le niveau du fonctionnement familial et du stress psychosocial étaient comparables au niveau de la population générale. Les enfants de familles dans lesquelles il y avait plus de problèmes familiaux, avaient plus de troubles du comportement et émotionnels.

En chapitre 7, les résultats des enfants avec la maladie de Hirschprung ou une malformation anorectale sont décrits. Les problèmes physiques les plus importants de ce groupe étaient les problèmes concernant la continence fécale. Les enfants avec une malformation anorectale avaient des problèmes plus graves avec la continence fécale que les enfants avec la maladie de Hirschprung. Pour les enfants qui avaient des malformations urogénitales, les problèmes de continence fécale étaient les plus graves. Dans ce groupe d'enfants avec la maladie de Hirschprung ou une malformation anorectale aussi, le quotient d'intelligence était plus bas que dans la population générale et plus d'enfants participaient à l'éducation spéciale. Deux fois plus d'enfants (30 %) que dans la population générale avaient des scores élevés sur le CBCL et le TRF. Il n'y avait pas de scores élevés pour dépression ni de scores plus bas pour l'image de soi. Il n'y avait pas de différences de fonctionnement psychosocial entre les enfants avec la maladie de Hirschprung et les enfants avec une malformation anorectale, pas plus qu'entre les enfants avec ou sans problèmes de continence fécale. Ces résultats étaient contraires à ce qui était prévu, parce qu'au préalable nous avions l'hypothèse que l'incontinence fécale causerait tant de problèmes que le niveau de fonctionnement psychosocial serait plus bas pour de tels enfants.

En chapitre 8, les résultats du groupe entier sont présentés. Les résultats du niveau de fonctionnement psychosocial des enfants, du fonctionnement familial et du fonctionnement des parents sont comparés à la population générale. Ensuite, le fonctionnement psychosocial des enfants est relié aux aspects de fonctionnement médicaux et familiaux. Pour cela, le 'disability-stress-coping' modèle de Wallander et Varni est employé. Dans ce modèle le fonctionnement psychosocial des enfants est relié aux facteurs de risque, tels que certains aspects de l'anomalie et le stress psychosocial, et aux facteurs protectrices, tels que les facteurs intrapersonnels et le fonctionnement familial ou psychosocial des parents.

Bien que 27 enfants avaient des problèmes physiques modérés, le fonctionnement physique des enfants peut être considéré comme bien. Avec l'exception de deux enfants avec de problèmes auditifs, les enfants n'avaient pas de limitations physiques graves. Le quotient d'intelligence moyen du groupe était de 8,5 points plus bas que la norme de 100 et la participation à l'éducation spéciale était quatre fois (17 %) plus fréquent que dans la population générale. Trente pour cent des enfants avait des scores élevés sur le CBCL et TRF, comparé à 15 % dans la population générale. Les scores des enfants concernant la dépression n'étaient pas plus hauts et concernant l'image de soi n'étaient pas plus bas que dans la population générale. Le fonctionnement familial, la relation conjugale et le fonctionnement psychosocial des parents étaient tous dans la portée normale. La relation entre les facteurs liés l'anomalie, tels que le diagnostic ou la gravité des problèmes physique au moment de l'étude d'un côté et le fonctionnement psychosocial des enfants de l'autre côté était limitée. Mais les enfants avec des anomalies majeures multiples avaient un quotient d'intelligence bien plus bas (QI moyen 76,9) que les enfants avec seulement une anomalie majeure (QI moyen 94,4). Le fonctionnement psychosocial était surtout relié au stress psychosocial qui était directement lié à l'anomalie congénitale, rapporté par les parents et les enfants, et au niveau du fonctionnement familial, rapporté par les parents. Ces deux facteurs ensemble expliquaient entre 17,1 % et 35,1 % de la variance dans le fonctionnement psychosocial des enfants. Le degré du stress lié à l'anomalie était indépendant de la nature de l'anomalie. Le modèle de Wallander et Varni fut un moyen important pour étudier les relations entre l'issue et les facteurs déterminants et ce modèle pouvait partiellement être confirmé dans cette étude.

Les résultats de cette étude mènent à quelques importantes conclusions concernant le fonctionnement psychosocial des enfants avec des anomalies congénitales abdominales majeures, lesquelles sont exposé dans chapitre 9. Notre étude a montré que des enfants avec des anomalies congénitales abdominales majeures ont un risque élevé de problèmes psychosociaux. Ces problèmes sont indépendants de la nature de l'anomalie, mais étaient surtout relié au stress psychosocial lié à l'anomalie et au fonctionnement familial. Apparemment, toutes les anomalies congénitales abdominales majeures causent autant de stress, que le fonctionnement psychosocial en est influencé négativement. Bien sur, le stress est différent pour chaque anomalie. Le stress lié

à une anomalie congénitale abdominale majeure peut se composer de problèmes de continence fécale, d'une cicatrice de l'opération ou de limitations de l'endurance physique causées par des problèmes respiratoires. Dans des recherches futures ces résultats doivent être incorporés. Il est important d'inclure toutes les anomalies congénitales abdominales majeures dans l'étude et non seulement un nombre de diagnostics limité.

De cette recherche quelques recommandations pour de futures recherches pourraient être données. Il nous semble nécessaire de projeter des études de suite d'une nature prospective et longitudinale, de mesurer plusieurs aspects du fonctionnement des enfants d'une manière standardisée, d'incorporer le jugement des parents aussi bien que celui des enfants et d'incorporer une plus grande diversité d'anomalies congénitales dans une telle étude. Coopération plus proche entre les divisions de chirurgie d'enfant, de pédiatrie, de psychologie médicale et de psychiatrie d'enfant est nécessaire. Pour la pratique clinique, il serait de grande importance de reconnaître que le groupe entier d'enfants ayant des anomalies congénitales abdominales majeures peuvent avoir de problèmes psychosociaux et qu'il faut être alerte à ces problèmes pendant les visites de suite à l'hôpital. Les enfants qui ont un risque élevé de problèmes psychosociaux, font partie de familles avec plus de problèmes et avec un plus haut niveau de stress lié aux anomalies. Des études de suite d'un tel groupe jusqu'à l'âge adulte sont nécessaires afin d'augmenter la compréhension du développement de pareils enfants.

Dankwoord

Een proefschrift is de vrucht van een paar jaar werk van veel meer dan één persoon en draagt tegelijkertijd een ontwikkeling in zich van vele jaren. Dit betekent dat mijn dank zich uitspreid over velen. Een aantal van degenen die ik dank verschuldigd ben wil ik hier noemen.

Het onderzoek waarop dit proefschrift is gebaseerd was niet mogelijk zonder de medewerking van vele kinderen en ouders. Zowel de kinderen en ouders die het onderwerp zijn van dit onderzoek, de kinderen met aangeboren darmafwijkingen, als de kinderen en ouders uit de vergelijkingsgroep, die feitelijk geheel belangenloos meewerkten.

Er zijn twee personen die een zeer grote bijdrage aan het onderzoek en aan dit proefschrift hebben geleverd en zonder wie dit ook werkelijk niet tot stand was gekomen. Annabel van Gijs, je hebt twee jaar lang in alle ijver en nauwgezetheid de dataverzameling en analyses voorbereid en mede uitgevoerd. Je grote waarde was dat je zoveel orde hebt aangebracht in de data, dat de chaos die ik vervolgens heb aangericht daar niet tegenop kon. Hans Koot, je was vanaf het begin betrokken bij de opzet en uitwerking van het onderzoek. Het is vrijwel onmogelijk te beschrijven hoeveel je voor mij hebt betekend de afgelopen jaren en ik wil dan maar vooral de hoop uitspreken dat ik in de toekomst nog vaak het voorrecht zal mogen hebben met jou samen te werken.

Het initiatief voor dit onderzoek is uitgegaan van Frank Verhulst en Jan Molenaar. Frank, je bent voor mij vooral een inspiratiebron geweest de afgelopen jaren, door de enorme werklust en consistentie waarmee je een onderzoekslijn op de afdeling hebt opgezet en uitgebouwd. Je belangstelling en de wezenlijke adviezen die je gaf, weliswaar vanaf enige afstand, waren voor mij van zeer grote waarde. Jan, je stond aan de wieg van dit onderzoek. Het tekent jouw brede visie op de geneeskunde en de kinderheeskunde in het bijzonder, dat je dit onderzoek hebt geïnitieerd en steeds met belangstelling gevolgd, ook na je emeritaat.

Eén van de meest plezierige aspecten van het werken als consultatief kinderpsychiater is de samenwerking met andere medische disciplines en wel in het bijzonder in het kader dit onderzoek met de afdeling kinderheeskunde van het SKZ. Frans Hazebroek, je was de afgelopen 4 jaar nauw betrokken bij de onderzoeken van de kinderen en later bij het voorbereiden van publicaties. Met een bijzondere rust, maar vooral met enthousiasme en warme klinische belangstelling heb je dat gedaan. Dick Tibboel, je leidde me lang geleden rond op de intensive care kinderheeskunde en daar maakte ik kennis met je gedrevenheid maar ook met je belangstelling voor de psychosociale aspecten van je werk. Richard Langemeijer, jij liet me in jouw dagelijkse praktijk zien wat de consequenties van een aangeboren darmafwijking kunnen zijn voor het leven van een kind. Alle andere leden van de staf van de afdeling kinderheeskunde en Carla Freund dank ik voor jullie bijdragen en vriendelijke steun die ik de loop van het onderzoek mocht ontvangen.

De overige leden van de kleine commissie wil ik danken voor de tijd en moeite die ze

namen om het proefschrift te lezen en te beoordelen. Jan Passchier, jij was bij de eerste besprekingen betrokken en je warme belangstelling in de loop van het onderzoek is zeer stimulerend voor me geweest. Herman van Engeland, je stond aan de wieg van mijn opleiding tot kinder- en jeugdpsychiater maar ook in de loop van de jaren bleef je geïnteresseerd en gaf me soms net een duwtje in de rug om door te gaan.

Onderzoek op de afdeling kinder- en jeugdpsychiatrie van het SKZ doe je niet alleen. In alle jaren heb ik me ook steeds een beetje AIO gevoeld en van velen van hen mocht ik steeds veel vriendschappelijkheid ondervinden. Enkelen van hen, Kuni Simis, Jeroen Heijmens Visser, en Jolande van der Valk wil ik succes wensen bij de voorbereiding op hun eindresultaat. Jan van der Ende, bij jou binnenlopen met een statistische vraag was nooit een probleem en altijd gaf je een helder en praktisch antwoord.

De afgelopen jaren heb ik onderzoek en kliniek kunnen combineren, waarvoor ik zeer dankbaar ben. En dus ook aan de 'klinische kant van de gang' zijn er mensen die ik wil noemen. Guy Berden je was een zeer prettige collega en onder jouw leiding konden ik en velen met mij op de polikliniek in een goede sfeer ons werk doen. Collega kinderpsychiaters, jullie waren steeds heel prettige collega's, met name Dirk de Raeymaecker, collega van het eerste uur, dank voor je enorme hulp bij de Franse vertaling van de samenvatting. In het bijzondere wil ik hen noemen met wie ik de afgelopen jaren nauw heb samengewerkt. Nel van Bodegom, met jou heb ik nu al 7 jaar met meer dan gewoon plezier samengewerkt. Voor je nauwgezetheid, je betrokkenheid en je vriendschap wil ik je hartelijk danken. De arts-assistenten, jullie gaven me de gelegenheid iets van mijn eigen kennis en ervaring over te dragen. Jullie soms prikkelende en kritische houding waren een belangrijke stimulans voor me. Alle andere collega's, ik wil jullie ik danken, omdat ook jullie het werken op de afdeling kinder- en jeugdpsychiatrie de afgelopen jaren tot een genoegen hebben gemaakt. Medewerkers van het secretariaat voor het plezier dat we samen kunnen hebben, voor het verdragen van mijn chaos en voor jullie bereidheid om steeds weer op mijn 'tussendoor' vragen in te gaan. Alle andere 'administratieve' werkers die altijd bereid waren mij met een of andere vraag terzijde te staan.

Elly Dekker voor het kritisch doorlezen en becommentariëren van enkele artikelen en vooral voor je vriendschap en die van Cees van zoveel jaren.

Pierre Collignon ontwierp de omslag van dit proefschrift. Pierre, je hebt dat met een luisterend oor en een observerende blik gedaan en je brengt op de omslag prachtig in beeld wat binnenin in woorden is beschreven.

Een in een proefschrift meestal onzichtbaar facet van iemands bestaan is zijn achtergrond en privéleven. Mijn ouders wil ik noemen voor wat ze me in zoveel jaren hebben gegeven en waarmee ze in mijn gedachten blijven voortleven. Mariet, Arie, Maria, Annemieke en Jan, ik kan alleen maar dankbaar zijn, dat jullie er zijn, dat het heerlijk is met jullie samen thuis of uit te zijn en dat geen belang ook niet van een proefschrift boven jullie uit kan stijgen.

Curriculum vitae

Nicolaas Hendricus Bouman werd geboren op 26 februari 1955 te Dordrecht. Hij groeide op in Kinderdijk met zicht op rivier, boezem en molens. In 1973 deed hij eindexamen gymnasium β aan het Christelijk Lyceum te Dordrecht.

Hij studeerde vanaf 1973 geneeskunde aan de Erasmus Universiteit Rotterdam waar hij in 1980 zijn artsexamen behaalde. Na zijn artsexamen studeerde hij een jaar filosofie eveneens aan de Erasmus Universiteit Rotterdam. Van 1981 tot 1982 deed hij zijn eerste schreden in de psychiatrie op de afdeling Beschut Wonen van psychiatrisch centrum St Bavo te Noordwijkerhout (Opleider J. Dröes). Medio 1982 startte hij zijn opleiding tot psychiater bij de RIAGG Zuid-Hollandse Eilanden te Spijkenisse (opleider J. Jenner). Van 1983 tot 1985 vervolgde hij zijn opleiding in psychiatrisch centrum Bloemendaal te Den Haag (Opleiders L. Joele en W. Nolen). Op de jeugdafdeling Laanzicht van Bloemendaal werd zijn belangstelling gewekt voor de kinder- en jeugdpsychiatrie (Opleider J. Duycx). Van eind 1985 tot eind 1986 deed hij zijn opleidingsjaar neurologie in het Diaconessenhuis te Heemstede (Opleider R. Groen). Van 1987 tot 1988 volgde hij de opleiding tot kinder- en jeugdpsychiater in het Academisch Ziekenhuis Utrecht (Opleider H. van Engeland). In dezelfde periode werkte hij part-time als psychiater in Groot Emaus te Ermelo.

Van 1989 tot 1991 werkte hij als kinder- en jeugdpsychiater in het Juliana Kinderziekenhuis te Den Haag, hoofd Th. Doreleijers, waar zijn interesse in de consultatieve kinder- en jeugdpsychiatrie ontstaan is. Sinds 1991 werkt hij op de afdeling kinder- en jeugdpsychiatrie van het Sophia Kinderziekenhuis, hoofd Prof. F. Verhulst. Het onderzoek waarop dit proefschrift is gebaseerd, werd de afgelopen jaren uitgevoerd op deze afdeling in samenwerking met de afdeling kinderheekunde van het Sophia Kinderziekenhuis

Samen met J van Weel richtte hij in 1990 het kinderconsultatieplatform op, tegenwoordig de Nederlandse Vereniging voor Psychosociale Consultatie in de Kindergeneeskunde, waar hij voorzitter van is.

Hij is getrouwd met Mariet Bieber en heeft 4 kinderen, Arie, Maria, Annemieke en Jan.