

**The value of ultrasonography as a screening
procedure for congenital dysplasia of the hip in
newborns**

(De waarde van echografie als screeningsonderzoek
naar congenitale heupdysplasie bij pasgeborenen)

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I.0 TERMS USED IN THIS STUDY

Congenital Dysplasia of the Hip:

A spectrum of disorders with a disturbed relationship between the femoral head and the acetabulum, that either exists at birth or may develop during the first months of life. The spectrum ranges from frank dislocation of the femoral head to a shallow acetabulum with a steep roof, less able to withstand weight bearing in later life.

Radiological Dysplasia:

Steepness of the acetabulum as seen on the radiogram. This has been quantified for age by a.o. Tönnis and Brunken. Depending on the severity, the femoral head may be inside the joint, subluxated or dislocated.

Ultrasound Dysplasia:

Steepness of the acetabulum as seen on the ultrasound image. This has been quantified by Graf (1986). If the child is over three months of age, the alpha angle (angle of bony cover) should not be less than sixty degrees for a hip to be classified as normal.

Physiological Immaturity:

A term introduced by Graf (1986) to indicate hips of infants up to three months of age, that have an angle of bony cover between fifty and sixty degrees.

Dislocation:

There is no contact between the femoral head and the acetabulum.

Subluxation:

There is a diminished contact between the femoral head and the acetabulum.

Dislocatable:

The femoral head is inside the acetabulum, but may be moved out of it by mild pressure.

Ultrasound Instability:

A very ill-defined term. In this study it is meant to signify the displacement of the femoral head inside the acetabulum that can sometimes be seen on the ultrasound screen in real-time, when the femoral head is put through its range of motion, or pressed upward and backward gently. The labrum of the acetabulum can be seen to displace with the femoral head.

Sliding of the hip:

A term sometimes used by clinical investigators of newborn hips, indicating a sensation of diminished stability during the clinical examination. This term should be abandoned as it causes confusion.

II.0 INTRODUCTION

II.1 Purpose of this study

Despite numerous publications on ultrasonography as a diagnostic tool for congenital dysplasia of the hip in infants up to one year of age, only a few recent studies have dealt with its role in screening unselected newborns, in addition to routine clinical examination (Berman and Klenerman 1986, Langer 1987, Benz-Bohm et al 1987, Castelcin and Sauter 1988, Exner 1988). With ultrasound it appeared that visualization of the newborn hip could be carried out on a large scale. This could perhaps elucidate the role that the shape of the neonatal acetabulum plays in the development of congenital dysplasia of the hip (Wynne-Davies 1970a,b).

Furthermore it could objectivate the tactile sensations of the examiner's hands in registering stability of the hip, or lack of it, as is manifest in the examination as proposed by Ortolani (1937), Barlow (1962) and others (see Chapter II.4)

A very important question remains to be answered, however:

What is the natural history of untreated 'abnormalities' at the newborn age that can only be detected by ultrasound ?

A pilot study carried out by the author, investigating this problem in 307 newborns (Castelein and Sauter 1988) has led to the present investigation.

The aim of the present prospective study, performed in an unselected population of newborns, was to answer the following questions:

1. Do hips with an insufficiently developed acetabulum at birth according to ultrasound criteria, that are normal according to clinical criteria, develop true, radiologically proven dysplasia in later life if they are not treated ?
2. Do hips, that are clinically normal at birth, but show lack of stability during the dynamic ultrasound examination, develop dysplasia ?

Since this study also involved a follow-up investigation of children beyond the age of 6 months in whom an X-ray of the pelvis was taken, an additional third question was asked:

3. What is the sensitivity and specificity of ultrasound at the age of at least 6 months, when compared with a pelvic radiogram ?

II.2. Definitions

Many terms have been used in the literature to designate the disorder that this study deals with.

Congenital dislocation, subluxation, preluxation and dysplasia of the hip have all been used more or less interchangeably by different authors, indicating different stages of a disturbed relationship between the femoral head and the acetabulum. Whether this disturbance exists already at birth in all cases or may develop during the first months of life remains a matter of dispute (as will be shown later), but has not influenced the semantics.

Hilgenreiner (1925) called the disorder an 'Entwicklungsverzögerung' rather than an

'Entwicklungshemmung'. This indicated, as Gill (1948) also described, a prenatal defect in the development of the structures of the hip, predisposing to degenerative changes in later life. The term 'Congenital Dysplasia of the Hip (CDH)' would be most suited to cover all the different manifestations of this abnormality.

In the course of this study, the term Congenital Dysplasia of the Hip or CDH will be used throughout. This is meant to imply both the hip joint that is actually subluxated, dislocated or dislocatable at birth, as well as the shallow and steep acetabulum that may lead to a faulty development of the joint later in life (Wynne-Davies 1970 a,b).

In ultrasonography, in addition to normal and dysplastic hips, the term 'physiologically immature hip' was introduced by Graf (1986). This indicates the hip with an acetabulum that appears steep on ultrasound, in newborns and infants up to three months of age. The alpha angle (angle of bony inclination) lies between fifty and sixty degrees. The osseous acetabular rim may be slightly rounded. These hips should be followed regularly for their development, since rapid deterioration may occur (Graf 1986, van Moppes and de Jong 1988b, Tönnis 1989).

II.3 Pathology of Congenital Dysplasia of the Hip

Watanabe (1974) and Coleman (1978) have summarized the embryological development of the human hip. The hip joints are formed during the embryological phase from one block of scleroderm. Four weeks after gestation, the embryo is approximately five millimeters long, and the primitive anlage of the extremities has formed. At six weeks, the embryo is one centimeter long. Femoral head and acetabulum have formed, although there is not yet a joint space. This area is filled with a large number of mesodermal cells, from which the joint cavity with its cartilage will develop. At eleven weeks, the joint cavity can be recognized. The diameter of the femoral head is approximately two millimeters. The acetabular labrum has formed, as well as the joint capsule. From this period on, gradual maturation of the existing structures takes place. Based on these data, no dislocation of the hip can possibly occur before the eleventh week of gestation.

Many studies have described the patho-anatomy of congenital dysplasia of the hip, they were summarized by Tönnis (1987). Stanisavljevic (1982) has shown that various forms of congenital dysplasia of the hip may exist at birth, ranging from true, established dislocation that may occur shortly after the twelfth week of intrauterine development, with very severe alterations of the proximal femur and acetabulum, to a mild disorder at birth that is characterized mainly by a slight enlargement of the capsule, a hyperelastic limbus, and a longer than normal ligamentum teres. The femoral head and the acetabulum have almost normal shapes. If untreated, these latter hips may undergo gradual deterioration under the influence of extension of the legs, and later weight bearing. Changes consist of a defect on the supero-posterior aspect of the acetabulum, a pushed-up limbus, increased femoral anteversion, and slight deformation of the femoral head. With extension of the hips the defect on the supero-posterior aspect of the acetabulum increases and becomes located more superiorly.

Later, often when the child starts to walk, the defect changes its position and becomes mostly superior and supero-anterior due to the increased anteversion of the femoral neck. Whereas the former type (also called teratological) shows a stable dislocation of the femoral head out of the acetabulum, that cannot be reduced due to the malformed and small acetabulum, with obstruction by the limbus in combination with a deformed femoral head, the latter type can be repositioned into the acetabulum at birth without force, as becomes manifest with the Ortolani manoeuvre (1937).

The most frequent congenital hip pathology at birth is the hip that can be or is dislocated out of the acetabulum with initially only slight anatomical changes.

This type is called the 'typical congenital dysplasia of the hip'. It is this type of hip pathology that the present study deals with.

II.4 History of screening for Congenital Dysplasia of the Hip in newborns

Whether an early diagnosis of congenital dysplasia of the hip is desired depends on the ability to treat at an early age, and the conviction that this is beneficial for the ultimate fate of the joint.

Thus the history of screening is the history of early treatment. It is at present generally accepted that the prognosis in congenital dysplasia of the hip depends directly upon the age at which treatment is commenced (Moore 1974). This has not always been the case, however.

Roser, already in 1879, stated that early diagnosis of congenital hip dysplasia was a prerequisite for its successful treatment.

He described the mechanism by which a dislocated hip can be reduced by abduction, and postulated that an adducted position *in utero* could cause the disorder. This adducted position would lead to pressure in the groin region. Due to anatomical differences between the male and female foetus, this pressure would feel more uncomfortable in the male than in the female, thus causing reflex motions to avoid this pressure in the male foetus more often than in the female. This could explain why congenital dysplasia of the hip was so much more prevalent in female children than in males, a fact certainly appreciated at that time.

Interestingly, Roser admits that he had never diagnosed a dysplasia before the age of one year, nor had he heard of anyone who had. He states, however, that in his opinion most cases could be healed by simple abduction treatment had the diagnosis been made at the newborn age. This theory, unfortunately, gained little acceptance at that time and was soon forgotten (Tönnis 1987), only to be rediscovered much later.

There was little interest in early diagnosis of congenital dysplasia of the hip at that time, because principles of conservative treatment were mainly dictated by Lorenz (1920), who maintained that the most favourable age for the commencement of treatment of the disorder was after the child had started to walk.

Due to this relatively late age at which treatment was started, usually considerable force was required to place the femoral head inside the acetabulum. Reduction was then maintained by immobilisation in a plaster of Paris, in full abduction.

In 1969, Salter et al demonstrated the catastrophic effect of this position on the vascularity of the femoral head, and this type of treatment is totally abandoned nowadays. Reduction during the third year of life according to Lorenz supposedly gave almost as good results as during the second. He considered an early diagnosis (before walking age) as rather uncertain. It is of interest to note that Lorenz also felt that treatment with bandages of younger children, who were not yet trained in their sanitary habits, was very hard to carry out for practical reasons, not so much on philosophical grounds. With the development of radiology it became possible to confirm his good primary results by röntgen examination.

Lorenz maintained that a joint was healed when the femoral head was protected from lateral and upward migration by any sort of acetabular roof, even if this roof provided less cover than normal. In spite of his great experience in both conservative and operative treatment of congenital dysplasia of the hip, his dogmatic attitude hindered the development of the philosophy of early diagnosis and treatment for a long time.

This lasted until the middle of the twentieth century, when, in evaluating the ultimate results of this type of treatment, Severin (1941a) studied a group of 330 patients treated between 1913-1932 according to Lorenz' principles.

In this accurate follow-up investigation, an anatomically normal joint was found in only 4.2% of the cases, whereas 73.7% showed some degree of subluxation or dislocation. This disappointing result marked the end of an era in which late treatment for congenital dysplasia of the hip was advocated.

Muller and Seddon (1953), however, were somewhat more optimistic about the results of late conservative treatment. They reported the late results of different types of treatment for congenital dysplasia of the hip. Only 264 patients of a total of 889 cases treated between 1891 and 1940, were actually traced back and examined. 1.7% Was treated during the first year of life, 30.7% between the ages of 3 and 5. 155 Patients were treated by conservative means only. In 80% of the unilateral cases so treated before the age of three, and in 67% of the bilateral cases the result was considered satisfactory, based on a clinical and radiological evaluation.

Surprisingly, even hips thus treated between the ages of three and seven, fared almost as well. Their relatively favourable results may be related to the fact that the scoring system employed gave three times as many points for the clinical assessment as for the radiological criteria. Since the majority of patients examined was below the age of forty, it may still have been too early for severe clinical problems despite an unsatisfactory radiological result (Severin 1941a).

Already in the beginning of the century, Le Damany (1912) had described eloquently how, in the newborn, gentle adduction of the hips, with some outward pressure of the thumb on the inner aspect of the thigh could lead to a 'secousse', or 'jerk' in cases where the hip was dislocatable. Subsequently, upon gently abducting the leg with some inward pressure of the middle finger of the examining hand on the greater trochanter, the femoral head could be brought back into the acetabulum over its posterior border.

This re-entry into the acetabulum was always associated with a 'ressaut' (ressauter = to jump over an obstacle).

Hilgenreiner (1925, 1935, 1938) was an advocate of early diagnosis and treatment. He

described a case of redislocation of a previously dislocated hip at the age of ten, eight years after supposedly successful reduction by Lorenz himself. This caused him to raise the suggestion that many more of Lorenz' patients might show similar results after a number of years.

He thus challenged the concept of treatment after walking age.

In 1935 he presented 157 cases treated before the age of one year, utilising his aluminum splint. He advised that treatment should ideally be started at the age of 4-6 months.

Putti (1929, 1933) presented his philosophy of early diagnosis and treatment to the joint meeting of the British Orthopaedic Association and American Orthopaedic Association in London in 1929, and again to the British Orthopaedic Association in 1932.

Based on his experience in Northern Italy, an area 'where these dislocations are so common', he presented 119 cases with an average age of four months (range 34 days-16 months) at the time of diagnosis, and the beginning of abduction treatment. He claimed complete cure in 113, or 94.9% of his cases. He strongly believed in confirming clinical suspicion by röntgenographic examination, also in the young child.

This even led him to advise routine X-ray examination **in every newborn !!** He was convinced that in order to improve the results of treatment for congenital dysplasia of the hip, the age at which treatment was begun had to be lowered, even 'if that be on the day of birth'.

Ortolani (1937) re-introduced this concept of early diagnosis in the newborn. He described the clinical test to which his name has been attached since. His 'segno dello scatto' has erroneously been translated into 'click phenomenon', suggesting a primarily audible sensation. 'Jerk sign', or 'ridge phenomenon' (Felländer et al 1970) seems a more appropriate description.

Ortolani stated that this clinical sign was decisive in the diagnosis in infants, since he considered X-ray examination to be uncertain before the epiphysial nucleus was ossified.

He urged early treatment by means of a pillow placed between the legs. His publications for a long time apparently were more noted by pediatricians than by orthopaedic surgeons, but gradually his work spread.

In Sweden, Palmén (1953, 1961, 1984), Selander (in von Rosen 1956), and von Rosen (1956) started screening programs. Hart (1949) introduced Ortolani's principles into the U.S.A., followed by Coleman (1956, 1978) and Harris et al (1960).

Barlow (1962) reported on his experience obtained in 9289 infants in Great Britain.

An important addition to the test named after Ortolani was given more or less independently by Palmén, Coleman and Barlow. They added a dislocation test which consists of essentially the opposite of the Ortolani manoeuvre. Some hips that were *in acetabulo* at birth could be made to dislocate by gently adducting the legs with the thighs flexed, and exerting outward pressure on the lesser trochanteric region by the examiners thumb.

By this time the value of early diagnosis of congenital dysplasia of the hip had gained world wide acceptance.

II.5 Results of screening programs in preventing late diagnosis of CDH:

'The value of any screening programme must be judged by its failures-the number of children who slip through the net and require treatment later'. (MacKenzie and Wilson 1981).

In the following chapter all cases of congenital dysplasia of the hip diagnosed after the newborn period (first two weeks of life) will be considered as 'failures' of the neonatal screening programs. This does not mean to answer the question whether such late diagnosed cases are actually missed at birth, or may develop later in the course of the first year of life. It simply means that screening in the newborn has not prevented late diagnosed cases, whatever their cause.

The basic premises that justify all screening efforts in newborns are:

1. that all cases that would develop manifest congenital dysplasia of the hip with its sequelae at a later age, can be discovered at birth,
2. that adequate treatment at this early age is easier and leads to a better result than later in life, and
3. that it should not lead to harmful overtreatment of normal babies.

Many conflicting reports about the effectiveness of screening newborns for congenital dysplasia of the hip have appeared in the literature, with striking differences between the best and the worst results.

Favourable Results

Von Rosen (1956, 1962) has been one of the strongest advocates of early diagnosis and treatment, and published some of the best results. He worked in Malmö, Sweden and was in the favourable position that more than 99% of all the children were born in the city's one and only obstetrical department. In 1952 P. Selander, a pediatrician, started to perform routine examinations of newborns including the hip joints, searching for the Ortolani phenomenon (von Rosen 1956).

From 1952-1960, forty cases of congenital dysplasia of the hip were discovered. One child escaped early (neonatal) diagnosis because it was transferred to another hospital as an emergency, and therefore was not examined properly. All clinical diagnosis were confirmed by radiography according to the method described by Andrén and von Rosen (1958). All cases were treated in an abduction splint **that the mother was not allowed to remove** until the hip was judged to be stable by the doctor. Splintage was usually discontinued after three months. No redislocations occurred after treatment according to these lines, and only three of the first eight patients failed to develop normal radiographs. Although the length of follow up was not stated specifically, it appears from the X-rays and the table shown in their paper (1956) that results were assessed after at least a number of years.

Fredensborg continued the work of von Rosen in Malmö (1976a,b). In the *Acta Paediatrica Scandinavica* of 1976 he described the further experience with the same regimen from 1956-1972, and reported similar favourable results. Of 58 759 newborns that were

examined within the first 4 days of life, 548 were diagnosed to have congenital dysplasia of the hip, or 0,93%. Surprisingly, the incidence differed over the period under observation, from less than 0.5% to almost 2%. Possibly this can be related to the retirement of the most experienced investigators (dr. Selander and dr. von Rosen), and the delegation of responsibility for the program to less experienced investigators. The diagnosis was missed in four cases (0.007%), all of these had been examined at birth and were considered to be normal. In three cases unilateral dysplasia was diagnosed after the child had started to walk and was noted to limp. In one case bilateral dysplasia was diagnosed at the age of five weeks at a Childrens Welfare Clinic. In retrospect, no explanation for these failures could be given.

It was noted that, based on the reported incidence for manifest congenital dysplasia of the hip in the Malmö area, approximately 5 times as many children were being treated for the supposed disorder than would have developed a manifest dysplasia had no screening taken place.

In the same year (Fredensborg, 1976b) described the late results of early treatment for congenital dysplasia of the hip in 111 children, with an average follow-up of ten years. Splintage in the von Rosen splint was started within the first few days of life, and maintained for an average of ten weeks. There were two failures of this initial treatment. One girl with a left sided dysplasia that was treated according to this protocol was shown to have a radiological dysplasia at the age of ten with a sloping acetabulum but a CE angle (Wiberg 1953) within the normal limits.

Another girl was similarly treated for bilateral dysplasia and showed slight enlargement of the femoral head and neck at the age of eight, possibly the result of avascular necrosis.

Follow-up examination consisted of a clinical and radiological evaluation, with a comparison to a control group of 222 normal children, studied for unrelated disorders. Both clinical features and radiological examination were similarly distributed in both groups. All joints except for the two previously mentioned were radiologically indistinguishable from normal.

Similar good results of early diagnosis and treatment were described by Palmén (1961, 1984), Coleman (1956), Barlow (1962, 1975), Finlay et al (1967), Stanisavljevic (1964), Mitchell (1972), Paterson (1976), Hansson et al (1983), Tredwell and Bell (1981), Dunn et al (1985), Miranda et al (1988) and Hadlow (1988).

Late-diagnosed cases had diminished after screening and early treatment were instituted, treated hips tended to become normal. That this occurred at the expense of treating a considerable number of cases that would have become normal anyway, was also noted by most authors.

The question of what constitutes normality remains, however. It is widely assumed that many dislocatable hips at birth become stable without treatment, but an 'unknown number of dysplastic hips never proceeds to dislocation but rather persists as dysplasia or subluxation into adult life and ultimately become painful due to the development of secondary osteoarthritis' (Coleman 1982 in *Congenital dislocation of the hip* p 187 ed M.O. Tachdjian).

Less favourable results

Other reports have challenged the effectiveness of screening newborns for congenital dysplasia of the hip, and its early treatment. Robertson, in the *Lancet* (1984) even called neonatal screening for congenital dysplasia of the hip 'a mess'.

Moore (1974) describes four reasons why the diagnosis may be missed at birth.

1. Inexperience of the examiner.
2. Technical difficulty of the examination.
3. Irreducibility of the dislocation.
4. 'Late' dislocation of a previously normal hip.

Most authors admit that it has been impossible to completely avoid the problems of late diagnosis.

Jones (1977), in a study that involved almost 30,000 live births in a population of 400,000 over a period of five years found 17 that escaped neonatal diagnosis. No less than 16 were diagnosed after walking age.

Mitchell (1972) found four dislocated hips after walking age, in a screening population of 31,961 births, or one in 8000 newborns. Eight treated cases out of a hundred luxations diagnosed at birth failed to respond to treatment by splintage. Two showed deformity of the femoral head, in five a surgical reduction was required at a later stage, and one needed a derotation osteotomy. All 126 dislocatable hips at birth responded well to this type of treatment. It was suggested that failure to obtain or maintain initial reduction was probably due to an infolded limbus, as was demonstrated by arthrography.

Mackenzie (1972) reported 1.12 missed cases per 1000 live births for a large screening service distributed over a number of hospitals in the North-East of Scotland, started in 1960. Before screening started 0.15% of live births needed an operation for late diagnosed congenital hip dysplasia. Between 1970-1979 this percentage rose to 0.18% (Mackenzie and Wilson 1981).

1.11 Cases were missed at the newborn examination per thousand live births, a figure almost identical to that reported in their earlier series. No improvement had been made in early diagnosis despite ten more years of experience.

Williamson (1972) of Northern Ireland also reported difficulties in organizing a screening service in a widespread community.

His study shows a rise in the rate of diagnosis of dislocation or dislocatability of the hip at birth, but no appreciable fall in the number of children diagnosed late with an established dislocation. Thus early treatment with splints failed to prevent established dislocation in 2.4% of the treated hips.

Weisman and Salama (1966), in a series of 16,841 newborns examined with the Ortolani test between 1962-1964 found seventy dislocated hips in forty-five newborns.

Forty of these, with sixty-five dislocated hips could be followed for at least thirteen months. In spite of early abduction treatment in a von Rosen splint or Frejka pillow (Frejka 1941), half of the patients required a prolonged treatment ranging from six to thirteen months. A number of children still had dislocated hips inside the splint at three months of age, and movements of the femoral head into and out of the acetabulum were taking place during movements of the child.

Bjerkreim (1974) of Norway found no decrease in the number of late diagnosed cases of congenital dysplasia of the hip between 1960-1970, despite the introduction of the Ortolani test in 1953. This number amounts to 0.2% of live births (or a total of 184 newborns), even in the city of Oslo where almost all babies were examined by experienced pediatricians. It appears that no sign of hip dysplasia was present at birth in these babies.

Of those treated immediately after birth with an abduction pillow (Frejka 1941) 72% required no further treatment.

Only 47.6% of the total group of 1,121 patients that received primary treatment had entirely normal clinical and radiographical findings at follow-up.

In an effort to define the problem more clearly, Wilkinson (1972) tried to discover a 'position of dislocation' that would be only common to newborns with the disorder that was searched for.

In his philosophy, the principle of intra-uterine leg folding is crucial for the development of the hip. He could show that, in breech born children, evidence of delayed leg folding during intra uterine development was present in 80% of newborns with hip displacement, as compared to 65% of normal babies born by the breech. This correlation of leg folding mechanism and dysplasia of the hip was less clear after vertex presentation. Out of a group of 6,272 live births, eleven (0.17%) developed persistent dysplasia, without a positive Ortolani sign at birth.

This focused attention on the so-called irreducible hip at birth, that appeared to have a more gloomy prognosis than the reducible hip. The former type would be difficult to diagnose at birth because of lack of reducibility, required for the Ortolani manoeuvre. Response to early splintage was poor in these hips.

Both groups seemed to share the same etiological factors, and were considered to be probably different degrees of the same deformity. Thus Wilkinson acknowledged that there was a group of hips that could not be diagnosed with the traditional examination as described by Ortolani and Barlow. In most studies these hips would present late, often after walking age.

In 1975, Wilkinson elaborated on this problem. He described three cases, two failing to respond to conservative treatment that was started right after birth, the third with a negative diagnosis at birth despite a manifest dysplasia at three months of age. At this time conservative treatment was instituted, that ultimately led to fragmentation of both hips due to ischaemic necrosis.

He concluded that possibly in some cases it would be better if the diagnosis of congenital dysplasia of the hip would not be made until after the first year of life. The result of surgical treatment in the second year could, in some instances, be better than the result of early conservative treatment in two of his cases.

Place et al (1978) reported that the incidence of late diagnosed cases of CDH in screened series had remained at a rate not very different from that reported before screening was started.

This assertion was also made by other authors (Catford et al 1982, Klingberg et al 1976, Wilkinson 1975, 1985). Robertson (1984) believes that 25-50% of dislocated or dislocatable hips are missed on neonatal examination. The incidence of surgery required for

the disorder was no different from that reported before neonatal screening was started in the 1950s.

Zadik (1983) also cast doubt upon the effectiveness of early diagnosis. He reported a patient with a normal newborn examination.

At six weeks X-rays of the hips, taken for abdominal complaints, were normal. However at 16 months a right sided CDH was discovered.

Ifeld et al (1986) also made a very interesting observation in fifteen patients. These patients were all examined at various stages in their development by very qualified doctors (6 were professors specialized in children's orthopaedics). In all cases, despite multiple normal physical examinations previously, an increased acetabular index, subluxation or dislocation of the hip was found at a later stage. It appeared from their data that 9 of these children were examined shortly after birth, although not necessarily on the day of birth. One case was probably an example of true 'delayed dysplasia' in the sense that the right hip of this child was normal on multiple radiograms until 31 months of age (these were taken for evaluation of the **left hip** that was treated for CDH by open reduction and *bilateral* abduction cast). At four years of age a dysplastic **right** acetabulum was discovered with subluxation of the right femoral head. No other abnormalities were found in this child. It was concluded that CDH may be discovered months or even years after previous normal physical examination of the hip.

Kliscic et al (1988) state that CDH is a dynamic developmental anomaly. According to his experience it has become clear in recent years that its prevention cannot rely fully on screening in newborns. He proposes the term 'developmental displacement of the hip' to emphasize the dynamics of the disorder.

Early and late diagnosed congenital dysplasia of the hip

The concept of different types of congenital dysplasia of the hip, one that could be diagnosed in the newborn, another that would present late in most cases, was further investigated by Wynne-Davies (1970 a). She studied 1,897 first degree relatives of 589 patients with CDH. Of these, 192 had presented early, usually with an Ortolani 'clunk' at birth, 397 were diagnosed late. She found a significantly shallower acetabulum in first degree relatives of late-diagnosed cases as compared with controls, whereas early-diagnosed cases did not show that significant difference. Neonatal cases and their first degree relatives more often showed generalized joint laxity, compared to the late-diagnosed cases. It appeared that there were two distinct etiological groups, one a group with primary acetabular dysplasia, the other with primary ligamentous laxity. In the group with the acetabular dysplasia, the femoral head was stable inside a shallow and steep acetabulum at birth, thus the diagnosis could not be made by the Ortolani or Barlow tests. Due to the muscular forces and later weight bearing, the femoral head would slide out of the acetabulum, leading to a late-diagnosed dislocation.

The group with the generalized ligamentous laxity **could** be diagnosed by means of the tests of Ortolani and Barlow.

In some cases, due to persistent malpositioning of the femoral head opposite the acetabulum, the development of both parts of the joint would be defective, with secondary dysplasia as the result.

Acetabular shape was considered to be inherited as a multiple gene system, according to Wynne-Davies (1970 b).

Walker (1971) also expressed the opinion that some hips that were clinically normal at birth could dislocate later.

Davies and Walker (1984) presented 10 children with a normal clinical examination at birth, who developed acetabular dysplasia later in life. Four hips eventually dislocated.

Cyvin (1977) in a well controlled study of 19,864 infants examined for hip instability no later than the second day of life (the vast majority by the author himself), found 44 children with late-diagnosed CDH. They were all registered as having stable hips at birth. From comparison with a group of 378 children with neonatal hip instability, he concluded that both groups represent separate clinical entities with a different etiology and pathogenesis.

Leck (1986) gave a critical epidemiological appraisal of screening for CDH. He concludes that the screening tests are not very accurate in detecting future cases of established CDH, since the latter are far less common than are positive tests.

However, the prevalence of late cases in nearly three-quarters of the screening studies was below the lower limit of the range for unscreened populations, indicating that treating neonatal instability prevents at least some cases of established CDH in early childhood.

In a recent study, Miranda et al (1988) come to the same conclusion. They calculate that, based on their material, only fourteen children (at most) required treatment for congenital dysplasia of the hip after screening was instituted, as opposed to seventy-nine had no neonatal screening been performed.

Is examination of the hips at the newborn age harmful ?

It has been suggested that examining the infant hip may damage the joint, especially the dislocation-provocation manoeuvre as described by Coleman, Palmén and Barlow (Cyvin 1977, Stanisvljevic 1964, Moore 1989).

Although it is easier to fracture the femur than to dislocate a normal hip at necropsy (Dunn 1969, Sommerville 1953), some hips with borderline stability may indeed be provoked to deteriorate by the dislocation manoeuvre. Also, simple extension of the newborn hip can lead to dislocation, as was shown by Salter (1968) in rabbits. If the examination in itself was harmful, this could explain the fact reported by some (Mackenzie and Wilson 1981, Wilkinson 1972, Moore 1989), that the number of late diagnosed cases of CDH in screened series exceeds the overall prevalence of the disorder in unscreened series.

II.6 Complications of early splintage

There could not be any opposition to abduction treatment of all newborns that are suspected of the disorder, indeed of all babies, even without any suspicion, if the abducted position of the legs was totally free of harm. This is not the case, however.

Felländer et al (1970) treated 69 hips in 42 newborns with an abduction splint for two to four months between 1958-1960. Osseous changes were found in the acetabulum and in the femoral epiphysis in 40%.

Mitchell (1972) described two cases of transient deformity of the hip after treatment in a splint. These (minor) changes were thought to be the result of avascular necrosis.

Wilkinson (1972) also reported interference with growth of the acetabulum and the femur due to splintage.

Mears (1974) even suggested that avascular necrosis due to treatment for CDH was so common that the newborn child should not be subjected to it.

Operative treatment carried less risk, and should be preferred.

In the *Lancet*, in an editorial on iatrogenic hip disease (1974), a similar warning was issued.

Fredensborg (1976b) reported one case with a possible avascular incident to the femoral head in a series of 111 patients treated in an abduction splint, in whom treatment started within the first few days of life. Fortunately, the eventual outcome was good.

Gore (1974) described 43 children treated in splints for unilateral CDH between the ages of one and sixteen months. A total of fifteen children developed avascular necrosis, five of them **bilateral** ! For these originally normal hips, the price of early splintage was very high.

Tönnis (1982), in a multi-center study, concluded that early treatment of CDH was absolutely essential, but should only employ the most atraumatic procedures. Thus, avoiding the position of maximum abduction and flexion as described by Lorenz (1920) in favour of the less extreme 'human position' as described by Salter et al (1969) was recommended.

According to Hierton and James (1968) 15-20 babies are splinted unnecessarily for every one that would develop manifest congenital dysplasia of the hip.

The fact that some cases appear to arise *de novo* may be explained by Wynne-Davies' (1970) theory of genetically determined acetabular dysplasia.

In summary, the incidence of congenital dysplasia of the hip at birth is reported to lie between 0.3 (Mitchell 1972) and 2% (Hierton and James 1968) of live births, depending on the series. Palmén (1984) reported even higher percentages in Scandinavian Laps, whereas Salter (1968) found extremely high percentages in North American Indians (12.3%). These figures are extraordinary, and are not reported from other areas. About 60% or more of hips that are dislocated or dislocatable at birth may heal uneventfully within one week without treatment (Barlow 1962).

The results of screening for congenital dysplasia of the hip vary in the literature from almost complete success in preventing late-diagnosed cases and the development of a normal joint after early treatment, to almost complete failure with as many late diagnosed cases or more as before screening started, and a high complication rate of early treatment. It must be noted, however, that the true prevalence of congenital dysplasia in the community remains obscure due the fact that a number of cases will continue to be discovered at later ages, when painful osteoarthritis develops.

Thus the estimated prevalence of the disorder in the unscreened populations may be too

low, and screening at birth may prevent some hips from going unnoticed until they would be detected at a later age due to the development of pain.

II.7 The role of standard radiography

In general, the role of radiography in the detection of congenital dysplasia of the hip in the newborn is limited. The newborn hip joint consists mainly of cartilage, and its true model is difficult to determine due to a lack of ossification of the acetabular edge. Also, the proximal femoral epiphysis is normally not yet ossified, so the exact relationship between the femoral head and the acetabulum is often unclear. The visualized bony elements represent only the centers of ossification but not the ultimate configuration of the joint. Furthermore, as described in the section on pathology, only minor changes in the shape of the hip joint exist at birth in most cases.

According to Coleman (1982) however, adequate X-ray examination taken at birth may still yield important information. He has given guidelines for the evaluation of the radiogram (1956) (Fig. 1).

Each determinant has a specific significance if it is abnormal.

The overall interpretation of the radiogram depends to a large extent on an evaluation of the shape and concavity of the acetabulum, possible lateral sclerosis of the acetabulum, lateroposition of the proximal femoral epiphysis and, at a later age, asymmetries in the ossification of the proximal femoral epiphysis. The most important single parameter, however, in the evaluation of a radiogram of an infant's pelvis is the acetabular angle or acetabular index according to Hilgenreiner (1925).

It indicates the slope of the acetabulum on the a.p. radiogram (Fig. 1).

In the newborn it is considered to be fairly unreliable, however, due to the lack of ossified landmarks (Caffey et al 1956, Coleman 1956).

Furthermore, its value will be substantially influenced by various positions of pelvic flexion, extension, or rotation, leading to asymmetrical projections.

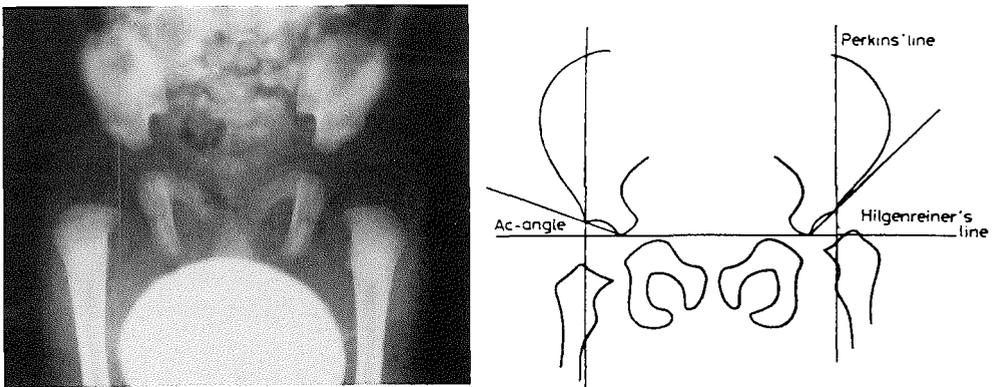


FIG. 1

Diagram and radiogram of an infant's pelvis. Schematic representation of Hilgenreiner's line, Perkins' line, and the Acetabular Index. Notice the lack of ossification.

Coleman (1956) emphasizes that the frequently occurring asymmetrical flexion contracture of the newborn hip must be taken into account when the infant is positioned for the radiograph.

For this reason Tönnis advises that an X-ray assistant holds the knees of the infant pointing symmetrically forward in slight flexion, corresponding to the physiologic flexion contracture (1987).

In spite of the disadvantages of plain radiography at this age, Coleman advises that when physical examination suggests hip abnormality, a single true anteroposterior view of the pelvis is mandatory. Not only may valuable information be obtained about the development of the hip joint if all possible pitfalls are taken into account, other disorders that mimic congenital dysplasia of the hip may be discovered as well if radiography is used. These include congenital short femur and coxa vara.

A normal X-ray at the newborn age does not mean that the hip is actually normal however, a view also held by Hensing (1982).

To eliminate the drawbacks of plain radiography in the newborn, Andrén and von Rosen (1958) described a technique to demonstrate the dislocated hip more reliably even in the very young child, following these principles:

1. The femora should be kept dislocated during the exposure of the film.
2. The diagnosis should not depend on angles between lines drawn from ill-defined points.
3. The diagnosis should not depend on uncertain measurements such as the one involving the upper end of the femoral metaphysis.
4. The results should be conclusive and leave no room for doubt.

In order to obtain these goals, the infant's lower extremities are held in forty-five degrees of abduction and an 'appreciable' degree of internal rotation. This manoeuvre forces the hip either into or out of the acetabulum, depending on the presence or absence of a dislocation. In normal hips, the line drawn through the femoral shaft will be directed towards the upper edge of the bony acetabular wall. In the presence of a dislocation, however, the line will point to the anterior superior iliac spine.

This technique was never used extensively in screening newborns for congenital dysplasia of the hip.

Palmén (1984) even states that in his opinion based on over thirty years of experience with neonatal screening for CDH, routine radiography in the newborn does not yield any additional information. In the older child, plain radiography yields more information thanks to the higher degree of ossification of the pelvis. In evaluating radiograms in infants, a number of possibly distorting influences must be taken into account.

Radiography provides a projection of a three-dimensional structure in one plane. Therefore, this projection is influenced by the position of the object relative to the plane of projection.

Both rotation of the pelvis around the longitudinal axis, as well as tilt of the pelvis around the transverse axis lead to changes in the acetabular index. These measurement errors may become significant, depending on the degree of rotation or tilt.

Tönnis and Brunken (1968) described their criteria to determine pelvic rotation around

the longitudinal body axis on the antero-posterior X-ray. For this purpose, the transverse diameter of the obturator foramina is measured (Fig. 2). If the quotient of these two measurements is between 0.56-1.8, the average error in measuring the left and right acetabular index should not exceed two degrees.

Visser and Konings (1982) have pointed out for adults with total hip arthroplasty that the angle of projection of the acetabulum is also influenced by the degree of its anteversion. Increased anteversion causes an increase of the angle.

Also, the acetabular index increases as the pelvis tilts backward, and decreases as the pelvis tilts forward (Tönnis and Brunken 1968).

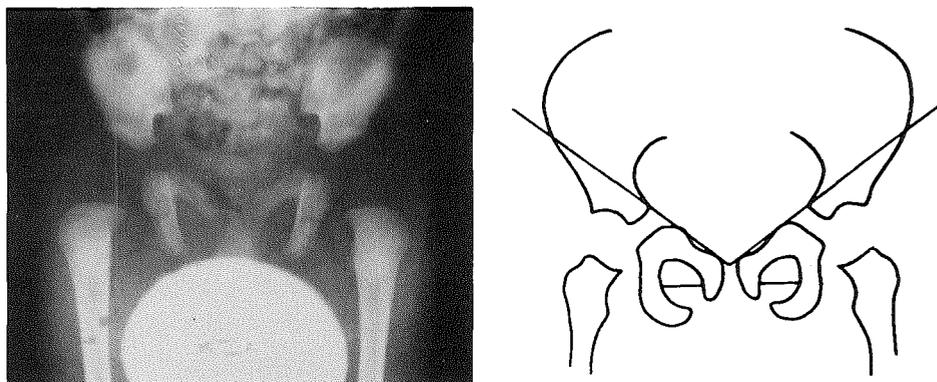


FIG. 2

Schematic representation of the diameter of the obturator foramina, and of the symphyseal-ischial angle. Two lines are drawn tangent to the highest point on each ischial bone, and meet at the point of the symphysis that projects farthest into the pelvis.

The symphyseal-ischial angle provides a guide to the determination of the degree of rotation around this transverse axis.

The angle is formed by two lines that are tangent to the highest point on each ischial bone, and meet at the point of the symphysis that projects farthest into the pelvic aperture (Fig. 2).

Normal symphyseal-ischial angles are listed below (Tönnis 1987):

1 - 6 months	_____	100° - 135°
7 - 12 months	_____	100° - 130°
1 - 2 years	_____	95° - 128°
2 - 3 years	_____	90° - 125°
3 - 5 years	_____	85° - 115°

In order to prevent a measuring error greater than 3 degrees, the symphyseal-ischial angle should be between these above mentioned values.

Normal values of the acetabular index

Normal values for the acetabular index depend on the age of the child, due to the varying degrees of ossification of the acetabular roof. Different values at birth are reported by different authors.

Kleinberg and Lieberman (1936) consider thirty degrees to be the upper limit of normal, but Caffey et al (1956) and Coleman (1956) demonstrated that at birth a wide variation in the size of the angles exists and that only a markedly elevated index may have some diagnostic significance (Coleman suggests over 40 degrees). Thirty degrees is given as a likely average value in the normal newborn.

As age increases, the measurement of the acetabular index becomes more reliable thanks to a better definition of the bony landmarks of the pelvis.

Mean normal values and standard deviations for various age ranges were given by a number of authors, most notably Tönnis and Brunken (1968), Massie and Howorth (1950), Wilkinson and Carter (1960) and Zippel (1971).

Although Krepler et al (1982) hold the opinion that the tables of Tönnis and Brunken show values for the acetabular index that may be too high, these tables are probably the ones that are used most frequently.

II.8 The role of arthrography

Arthrography enables the visualization of non ossified parts of the joint.

It is unnecessary to inject a contrast medium into the joint to demonstrate a high dislocation of the hip. This will be sufficiently evident from clinical examination and standard radiographs. Nor does it play any role in screening newborns.

But in order to obtain information about the factors that obstruct a concentric reduction of a dislocated or subluxated hip, and in planning treatment of recalcitrant cases that resist gradual reduction by mild methods, arthrography is indispensable.

The factors that may obstruct reduction include a stretched or constricted capsule, an interposed iliopsoas tendon, an elongated, enlarged or broadened ligamentum teres, an interposed labrum acetabulare, a hypertrophied pulvinar in the depth of the acetabulum, or a incongruency between the femoral head and the acetabulum.

Technique of the examination

In a very young child, general anaesthesia is preferred, in the older and very cooperative patient, local anaesthesia may be used.

There are basically four standard routes to enter the hip joint (Grech 1977). The joint can be entered from above, from below underneath the adductor tendons, from anterior, or from lateral. In all techniques the joint cavity is best entered by rotating the femoral head gently against the needle (Hughes 1982).

Tönnis, in his extremely comprehensive book on Congenital Dislocation and Dysplasia of the Hip (1987) discusses the history of arthrography. He feels that arthrography is essential in obtaining a truly atraumatic and individualized reduction of a dislocated hip,

because the soft tissue structures such as an interposed labrum, a hypertrophied pulvinar or a constricted capsule pose the greatest obstacle to anatomical reduction and are the cause of avascular necrosis.

Interpretation of the arthrogram

Mitchell (1963) and Severin (1939, 1941a,b, 1950) have performed invaluable work in the interpretation of the arthrographic findings in the normal and in the dysplastic hip. Severin concludes that the soft tissue interposition that is encountered in a dislocation of the hip is often gradually overcome after reduction. These interposed structures do not justify an open reposition *per se*, although his success rate in the completely dislocated hip was only 33 percent.

Tönnis (amongst others) has developed a system of grading dislocations based on arthrography as well as the results of reduction (1987). In grading the dislocation, a morphological description of the labrum acetabulare is combined with the position of the femoral head.

Four grades of dislocation are recognized.

Grade I: the cartilaginous head of the femur is laterally displaced by no more than 2/3 of its width; the labrum is everted and still covers the head.

Grade II: the femoral head is laterally displaced by more than 2/3 of its width but has not crossed the cartilaginous rim by more than 1/3 of its height in the vertical direction.

- a. The labrum is thinned, everted and still covers the head.
- b. The labrum is short, rounded, folded or deformed.

Grade III: The femoral head is displaced upward by more than 1/3 of its height relative to the cartilaginous rim of the acetabulum.

- a. The labrum is thinned, everted and still covers the head.
- b. The labrum is short, rounded, mildly inverted and deformed.

Grade IV: The femoral head is completely dislocated and is separated from the acetabulum by the labrum or the constricted capsule.

- a. The labrum hangs vertically, usually accompanied by the infolded capsule.
- b. A large labrum is inverted into the acetabulum and obstructs reduction.

It is essential that the radiographs are taken with the hip precisely in the neutral position.

In grading the quality of the reduction, the hip is flexed 110 degrees and abducted 50 degrees.

Three grades of reduction can be recognized on the arthrogram:

Grade I: The femoral head is fully reduced and is well approximated to the ischial part of the acetabulum.

Grade II: The femoral head is below the labrum but is somewhat lateralized due to constriction of the capsule and encroachment of the superior and inferior labrum and transverse ligament.

Grade III: The femoral head is not below the labrum and is outside the acetabulum. The labrum is inverted, or there is marked constriction of the capsule.

II.9 The role of computer tomography

Computed tomography can supply useful information about the anatomy of the hip joint, most notably in the transverse plane. Cartilaginous parts of the joint do not produce an image on the CTscan, so in the neonate the same objections as for routine radiography hold true. A contrast medium can be injected to overcome this obstacle, analogous to standard arthrography.

If enough of the femoral head is ossified, or a contrast medium is used, CTscanning may show the degree of coverage of the femoral head, the depth of the acetabulum, the congruity of the articular surfaces and acetabular and femoral anteversion.

The more combined anteversion of the acetabulum and the femoral neck exists, the less stable the joint will be.

Visser and Jonkers (1980) and Visser et al (1982) have described the so called 'instability index' on the CTscan, this index had previously been measured on cadavers (McKibbin 1970) and consists of the sum of the acetabular and femoral anteversion.

CT scanning should be reserved for those cases in which the other radiographic studies do not yield adequate information.

II.10 The role of ultrasonography

A number of studies has recently emerged, demonstrating the usefulness of ultrasound techniques in visualizing the infant hip joint, clearly initiated by dr. Graf of Austria (Berman et al 1986, Berman & Klenerman 1986, Boal & Schwenkter 1985, Clarke 1986, van Engelshoven et al 1988, Exner 1988, Graf 1980, 1981, 1982, 1982, 1983a,b,c, 1984, 1986, Harcke et al 1984, Morin et al 1985, Novick et al 1983, Pfeil et al 1987, Sanders et al 1988, Schuler 1984, 1987, Szöke et al 1988, Zick et al 1987, Zieger et al 1986, 1986, Zieger 1986, Zieger & Schultz 1987).

Ultrasound has the advantages over radiological imaging techniques that it does not employ ionizing radiation, that both bony structures and soft tissue can be visualized, and that dynamic examination may be performed in 'real-time'.

Although Kramps and Lenschow (1979) already mentioned the possibility of visualizing the cartilaginous acetabular labrum of the hip, the main impetus to the development of ultrasonography as a diagnostic aid in congenital dysplasia of the hip was undoubtedly given by Graf. He uses a lateral approach to the hip, placing the transducer on the region of the greater trochanter. This leads to an image of the investigated hip in the frontal plane. Others (Harcke et al 1984, Boal and Schwenkter 1985) have added an image in the transverse plane, to allow for three dimensional assessment.

Graf has given detailed accounts on the ultrasound anatomy of the infant hip, using waterbath experiments and anatomical dissections (1981, 1982). He found that hyaline cartilage (the unossified femoral head, the triradiate cartilage and the cartilaginous part of the acetabulum) does not produce detectable echoes, whereas the labrum acetabulare that consists of fibrous cartilage is echogenic. Bone produces strong echos (Fig. 3).

The size of the cartilaginous acetabulum can be inferred by the size of the gap between the echogenic labrum and the echogenic innominate bone.

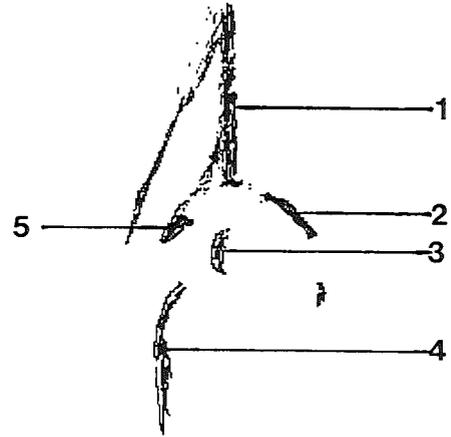


FIG. 3

Ultrasound image of an infant's hip. A very dark picture was produced on purpose to depict only truly echogenic structures. Normally the cartilage of the femoral head shows a few echos. 1: lateral aspect of the os ileum, 2: bony roof of the acetabulum, 3: ossific nucleus of the femoral head, 4: lateral aspect of the femoral metaphysis, 5: labrum acetabulare.

The femoral epiphysis becomes echogenic as it ossifies, and may be visible on ultrasound before its radiological appearance (Graf 1983c, Schuler & Rossak 1984).

Graf (1983a,b,c, 1986) has developed a classification, based on the ultrasound morphology of the bony and cartilaginous acetabulum in the frontal plane. Depth and slope of the acetabulum as well as the appearance of its bony and cartilaginous edge, determine if a hip is well formed, dysplastic, subluxated or dislocated (Fig. 4, 5, 6, 7).

Since ultrasound provides an image of one slice of the pelvis at a time, whereas plain radiography shows a cumulative picture of all different structures that are superimposed on top of each other, in ultrasonography the problem of finding a representative plane exists. This has also been emphasized by Graf (1986), who defines the standard plane as that plane in which the projection of the iliac bone is a straight line. If a slice further ventrally is chosen, the projected iliac bone curves outward, the projection of a more dorsal slice shows an inward curvature (Fig. 8, 9, 10).

For quantitative measurement Graf (1986) introduced two angles, alpha and beta, defined by three measurement lines (Fig. 11). The Baseline or Pelvic Wall Line is the line

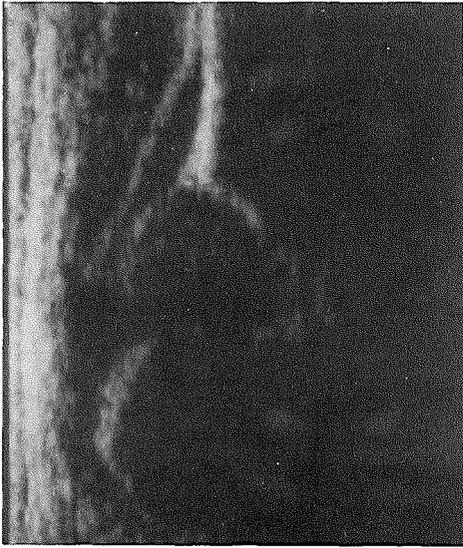


FIG. 4
Ultrasound image of a normal hip at birth.

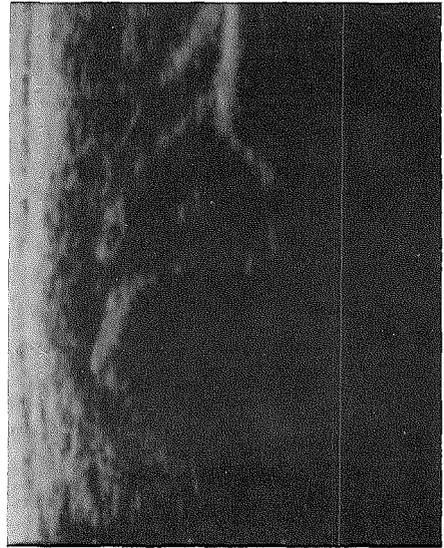


FIG. 5
Ultrasound image of a 'dysplastic' hip. The bony acetabulum is steep, its edge rounded.



FIG. 6
Ultrasound image of a subluxated hip. The femoral head is displaced proximally and laterally.

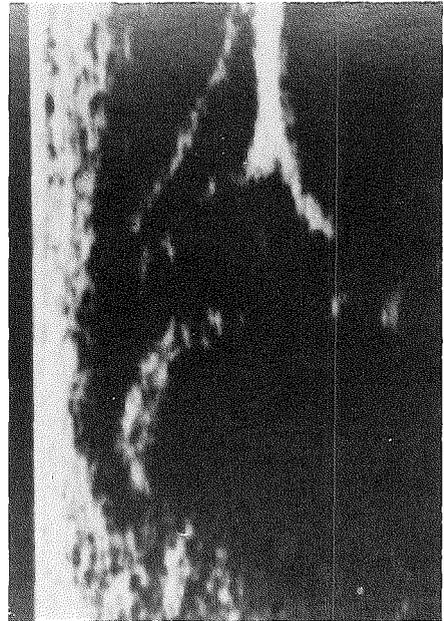


FIG. 7
Ultrasound image of a completely dislocate hip. The femoral head abuts against the lateral pelvic wall. No true acetabular cavity can be seen.

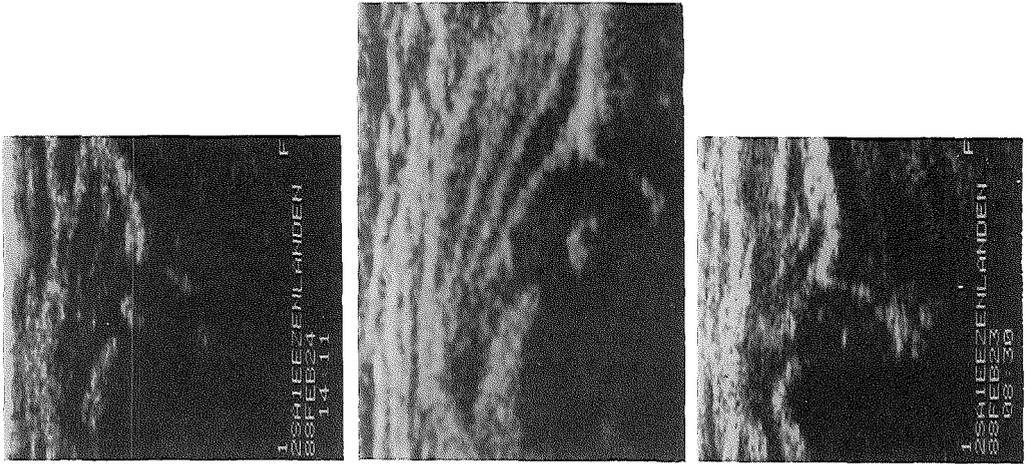


FIG. 8
Ultrasound image of a plane located ventrally through the hip. The echo of the os ilium curves outward. This image was obtained with a Pie Medical Scanner 450 Ortho, and reproduced with a Mitsubishi Videoprinter P60E.

FIG. 9
Ultrasound image of the standard plane through the hip. The os ilium is a straight line.

FIG. 10
Ultrasound image of a dorsal plane through the hip. The os ilium curves inward. This image was also obtained with a Pie Medical Scanner 450 Ortho, and reproduced with a Mitsubishi Videoprinter P60E.

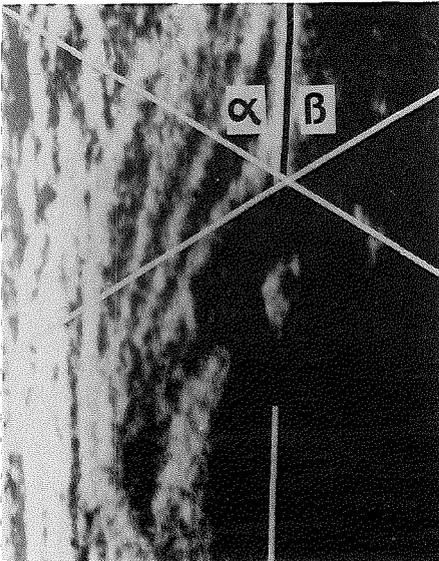


FIG. 11
Angle alpha and beta. Angle alpha corresponds to the inclination of the bony acetabulum, angle beta to the cartilaginous acetabulum.

that extends from the junction of the perichondrium of the cartilaginous acetabulum on the ilium tangent to the lateral wall of the ilium. The Bony Roof Line is the line that connects the point where the iliac part of the bony acetabulum ends, and the triradiate cartilage starts, to the bony rim of the acetabulum. The Cartilage Roof Line is the line that connects the superior bony rim of the acetabulum with the body of the labrum.

Angle alpha corresponds to the acclivity of the bony acetabulum, angle bêta to the inclination of the cartilaginous acetabulum.

Based on these lines and angles, four main hip types are recognized by Graf, and a number of sub-types (based on Graf in Tönnis 1987):

Type I. This represents the normal, fully mature hip, with either a sharply edged ossified acetabular roof or slight rounding and good coverage of the femoral head. The inclination of the bony acetabulum as expressed in angle alpha should not be less than sixty degrees in this hip type.

Depending on the shape of the cartilaginous acetabulum, hips can be classified as *Ia* in which the roof cartilage projects well laterally and distally over the cartilaginous head. The inclination of the cartilaginous roof (angle bêta) does not exceed fifty-five degrees.

Type Ib has a shorter cartilaginous roof, its bêta angle is greater than fifty-five degrees.

Type II: Well contained femoral head, the majority of the coverage is provided by the soft and deformable cartilage. There is roundness of the superior bony rim with a wide cartilaginous roof. This is associated with a delay in ossification. Angle alpha is between forty-four and sixty degrees, angle bêta is between fifty-five and seventy-seven degrees. In the infant below the age of three months, an alpha angle between fifty and sixty is called type *Ila*, and should be followed at regular intervals ('physiologically immature'). The same situation in the infant that is older than three months is called abnormal (type *Iib/c/d* depending on the alpha angle) and warrants treatment.

Type III. This represents further deterioration of the hip. The bony acetabulum becomes steeper, resulting in a lower alpha value ($<44^{\circ}$). The soft cartilaginous acetabulum is displaced upward. Depending on the echogenicity of the cartilaginous part of the acetabulum Graf subdivides into type *IIIa* and *IIIb*. The hyaline cartilage of the acetabular roof should normally be hypoechoic, type *IIIa*. In type *IIIb* pressure on the cartilaginous roof has led to a histological change. Hyaline cartilage is replaced by fibrous cartilage which shows as an echodense structure.

Type IV. Even further displacement, corresponding to a high hip dislocation. The dislocated femoral head and acetabulum cannot both be visualized in the same ultrasound cut, because they are no longer in the same plane.

The ultrasound image is not judged primarily on these angle measurements. Graf states that the overall impression of the ultrasound image, including the dynamic examination, is most important in the evaluation of a hip, the angle measurements merely provide for a 'fine-tuning' of the diagnosis. This is confirmed by Zieger (1986). Much emphasis is placed on the appearance of the bony and cartilaginous structures of the acetabulum, and whether the acetabular edge is sharp or rounded.

II.11 Possible future developments

Magnetic Resonance Imaging (MRI) has recently been described as a valuable adjunct to other available techniques in evaluating congenital dysplasia of the hip (Toby et al 1985, Bos et al 1988). MRI has the clear advantage of avoiding radiation exposure while providing a clear image of bony, cartilaginous as well as soft tissue structures such as muscles and ligaments without the need for a contrast medium. An important disadvantage is that the examination takes approximately 25 minutes, thus the (very young) child has to be sedated or even brought under general anaesthesia.

Although beautiful anatomical images may be obtained, its use will most likely be restricted to recalcitrant cases of congenital dislocation of the hip. At present its routine use is prohibited by the fact that only relatively few specialized institutes have these facilities, and by its cost and the need for sedation or anaesthesia.

III.0 SOME RELEVANT PHYSICAL PROPERTIES OF ULTRASONOGRAPHY

Ultrasound waves are waves of pressure, velocity and density that are propagated in a material element with a frequency greater than that of audible sound.

Ultrasound is a mechanical wave phenomenon; by definition it consists of sound waves inaudible to the human ear, i.e. exceeding 18,000-20,000 vibrations per second, or Hertz (Hz).

The usual application in medical diagnostics consists of frequencies above one million Herz, or Megahertz.

The industrial production of ultrasound is based on the piezzo-electrical effect. Synthetic crystals known as transducers transform the electrical energy into mechanical energy and vice versa. The frequency of the ultrasound waves depends on the mechanical characteristics of the crystal used.

Ultrasound can be employed in medical diagnostics, due to the fact that high frequency ultrasound possesses beam properties that are subject to the general laws of radiation.

Ultrasound can be directed into the body as an almost parallel beam, depending on the frequency that is used. The higher the frequency, the more parallel the ultrasound beam.

When it encounters an interface between tissues with different acoustic impedance, part of the ultrasound is reflected against the surface, thus giving rise to so called echos. The echos are received by the crystal, and re-transformed into an electrical signal.

Similar to audible sound waves, ultrasound requires a medium through which it can propagate. This medium can be gas, fluid or solid body.

As with all wave movements, ultrasound waves transport energy, that could potentially be harmful. During the propagation of ultrasound, tissue structures on a molecular level and a cellular level are brought into vibration.

The structure of the tissue (its mass and elasticity) are important parameters in determining the possible biological effects.

Ultrasound is non-ionising, has no mutagenic capacity and its effects are not cumulative. Cells that are damaged may either recover fully, or will die, depending on the dose that they receive. Ultrasound thus has an 'all or nothing' effect on the cell itself, but not on its 'offspring' (Williams 1983).

In general, the physical effects that ultrasound may theoretically have on tissue may be divided into thermic phenomena (development of heat) and non-thermic phenomena (radiation force, cavitation, micro current).

Development of Heat

Absorbtion causes development of heat due to the transformation of ultrasound energy. The absorbtion increases with the ultrasound frequency as well as with its intensity.

Biological effects can be expected if the temperature in the tissues rises above ± 42 degrees Celsius. Since the intensity of the ultrasound decreases with the depth inside the tissue, the development of heat is greatest on the surface. Furthermore, increase in temperature depends on the vascularity of the tissue.

In experimental animals that were subjected to diagnostic ultrasound investigations, no significant rise in temperature has ever been found (Lele 1982, Nyborg 1985).

Radiation Force

Ultrasound exerts a force on the border between two tissue planes. This force may lead to a biological effect. This radiation force has led to certain effects on suspensions of cells, such as stasis and aggregation (Dyson et al 1979).

Cavitation

Cavitation may take place if microscopically small gas bubbles start to oscillate under the influence of the ultrasound wave.

These bubbles can increase to a constant size (stable cavitation), or become unstable and collapse (transient cavitation). Both types produce mechanical and hydrodynamic forces that may damage biological material. The acoustic intensity required for transient cavitation to take place, is determined by the size of the gas bubbles. The higher the ultrasound frequency, the higher this threshold becomes.

Transient cavitation does not occur in vivo as long as the peak intensity of the ultrasound remains below 1500 W/cm^2 (Lele 1978).

Williams (1983) has shown that stable cavitation does not take place in blood, not even when small gas bubbles are injected.

Acoustic Microcurrent

An ultrasound wave exerts a radiation force on objects in a fluid. The cells in blood therefore concentrate around the areas of lowest local pressure (stasis). This has been noted to occur in vivo (Dyson et al 1979).

A second cause of microcurrent is the radiation torque that causes rotation of cells and even of intracellular structures.

The subsequent friction may damage the cell or its contents.

A third cause of microcurrent is stable cavitation. The gas bubbles that oscillate with high frequencies may damage the cells.

These phenomena except for stable cavitation have occurred in vitro, using continuous wave ultrasound (Doppler) and high intensities. There is no evidence that microcurrent is of significance when diagnostic intensities are used (Kremkau 1983).

Krizan and Williams (1973) have shown that a combination of the aforementioned processes may exert a biological effect.

In clinical practice, ultrasound is emitted in a pulsed manner. The pulse length is one microsecond or shorter followed by an interval of one millisecond. This means that during an investigation that lasts fifteen minutes, the tissues are only exposed to ultrasound during one second at most.

According to the American Institute for Ultrasound in Medicine (AIUM) 1982 statement, there is no evidence that exposure of mammalian tissues to diagnostic ultrasound in the frequency range of several Megahertz leads to biological effects.

IV.0 PATIENTS AND METHODS

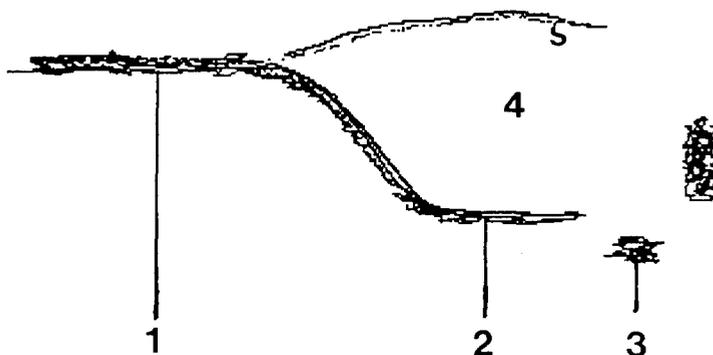
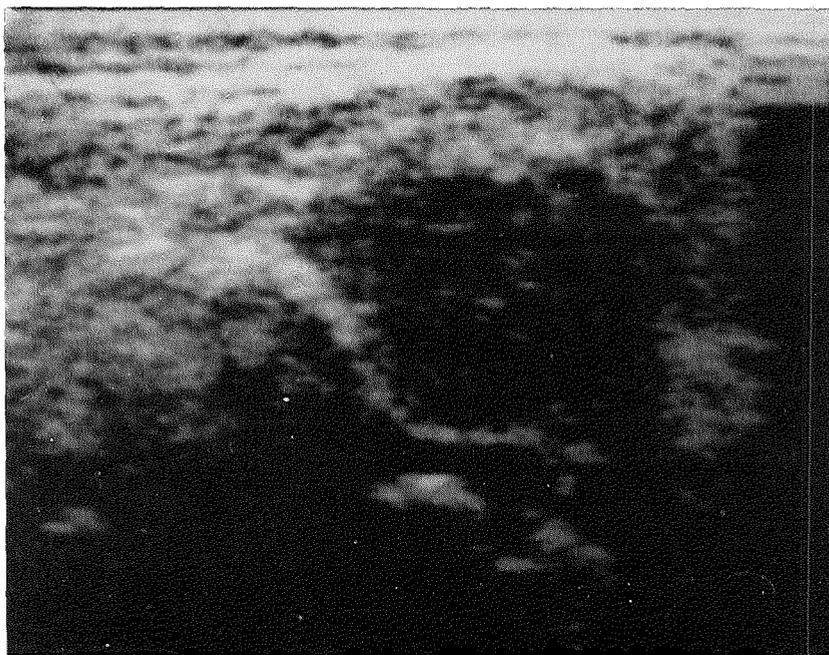


FIG. 12

Transverse cut through the hip. The image is comparable with a CT image. 1: pelvis, 2: bony acetabulum, 3: triradiate cartilage, 4: femoral head, no ossific nucleus present.

A total of 357 unselected newborns was examined according to a prospective, double blind protocol in the course of this study.

Parents were informed about the nature of this study, and asked if they wanted to participate. Relevant data about birth mechanism, birth order and family history were obtained from the mother. All newborns were examined independently by two investigators, as soon after birth as possible.

Clinical examination consisted of the reposition and dislocation manoeuvre described

in the introduction section (chapter II.4) that will be subsequently referred to as the Ortolani and Barlow tests.

The clinical part of the study was carried out by investigator I, an orthopaedic surgeon with experience in pediatric orthopaedics. Without knowledge of the results of the clinical examination, both hips of the newborn were subsequently scanned by the author of this study, investigator II. Hips were scanned with the newborn in the lateral position. No positioning device was used, the newborn was simply put on its side in its cot. A Philips SDR 1500 with 6 Mhz linear array transducer (type LA 6060) was used for this purpose.

An image was obtained in the frontal (coronal) plane, as described by Graf (1983a,b,c, 1986) (Fig. 4).

In addition, an image in the transverse plane as described by Harcke et al (1984) was obtained (Fig. 12).

In both planes a careful search for the most representative image was made, and of each hip at least two images in each plane were recorded for evaluation and measurement. Images were recorded with a 3M videoprinter (Dry Silver Imager, type 2 XR-I).

The ultrasound image was evaluated for a number of criteria. Overall impression was described with respect to coverage of the femoral head. The configuration of the bony acetabular corner (sharp or rounded) was noted, and three angles were measured. Angle alpha and beta as described by Graf were measured, in addition an angle gamma was introduced. This angle described the total coverage of the femoral head by the whole bony and cartilaginous acetabulum.

The gamma angle is formed by Graf's Base Line along the innominate bone and a line that connects the cranial border of the triradiate cartilage with the tip of the cartilaginous acetabulum, where it is joined by the labrum (Fig. 13).

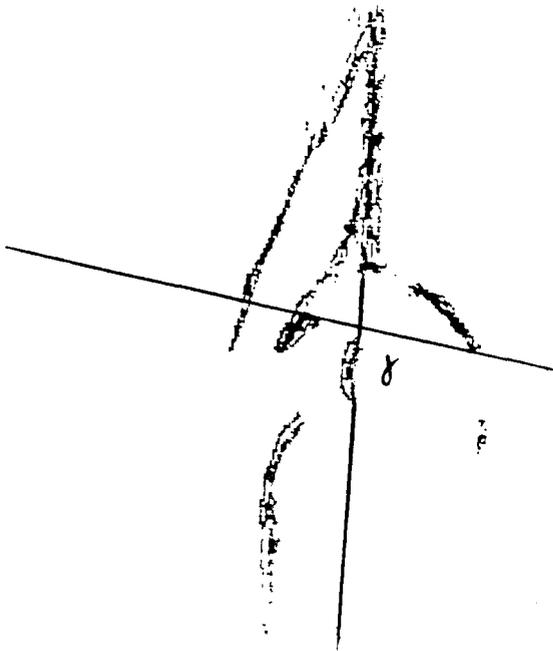


FIG. 13

Angle gamma. The angle is formed by the Base Line along the lateral aspect of the os ilium, and a line from the cranial border of the triradiate cartilage to the cranial border of the labrum acetabulare.

Under ultrasound control, a dynamic examination was performed. First the child was allowed to move its legs freely, and changes in the relationship between the femoral head and the acetabulum were noted. Subsequently the leg was gently moved through its range of motion. Finally, a gentle effort was made to push the leg upward, and move it from ventral to dorsal and back.

All data were entered on a computer worksheet, for computer processing in a personal computer.

The question of normality:

Clinical examination was considered to be normal if the hip was not dislocated, nor dislocatable (negative Ortolani and Barlow tests).

For the ultrasound examination, the most strict definition of normality was used.

The image was considered normal if:

1. The alpha angle according to Graf was at least sixty degrees.
2. No abnormal motion could be detected in the frontal or transverse plane.

Using sixty degrees as a lower limit for a hip to be considered normal means that, for the purpose of this study, type IIa hips according to Graf (1986), also called 'physiologically immature hips', were included in the potentially abnormal group. No treatment was instituted, but these hips were reexamined after an interval of at least six months.

Follow-up examination

Hips that showed ultrasound abnormalities according to investigator II (the author), but were considered normal on clinical examination by investigator I, were re-examined after an interval of at least six months. Directly after birth, the parents had been informed about the fact that standard clinical examination had not yielded any disorder, but that the ultrasound examination showed a possible delay in the development of the hip, that warranted another examination at a later stage. It was also explained that, although we had obtained experience with the method in over two thousand investigations, little was known about its relevance as a screening method in newborns.

Some infants were seen at a much later age, usually due to negligence or misunderstanding by the parents. This was felt to be undesirable, because in case of abnormality of the hip, too much valuable time had elapsed for effective treatment to be instituted. An effort was made to urge these parents to come back at the proposed time by means of telephone calls and letters, both to the home address and the family doctor.

At follow-up it was recorded whether the infant had begun to stand up or even walk (a number of children that were seen late actually had) and whether any disturbances in its development had been noted. The infant was examined clinically by the author, asymmetries in its movements were noted as well as asymmetrical abduction due to contractures.

A sonographic examination was performed along the lines described above. Furthermore, of all these children that were seen back standard antero-posterior radiograms of the hips were obtained. These radiograms were judged by an independent radiologist, with

experience in pediatric radiography, who did not know the results of the clinical nor of the ultrasound examination. Hips were classified based on their overall radiological appearance, and on the measurement of the acetabular index as definitely normal, dysplastic or borderline.

Definitely normal meant a well formed joint with an acetabular index lower than or equal to the normal values for age as given in the tables of Tönnis and Brunken (1968).

Borderline meant a hip with an acetabular index less than one standard deviation above the mean for age.

Dysplastic meant an acetabular index at least one standard deviation above the mean for age. Within this group a further distinction was made for hips with an acetabular index of more than two standard deviations above the mean for age.

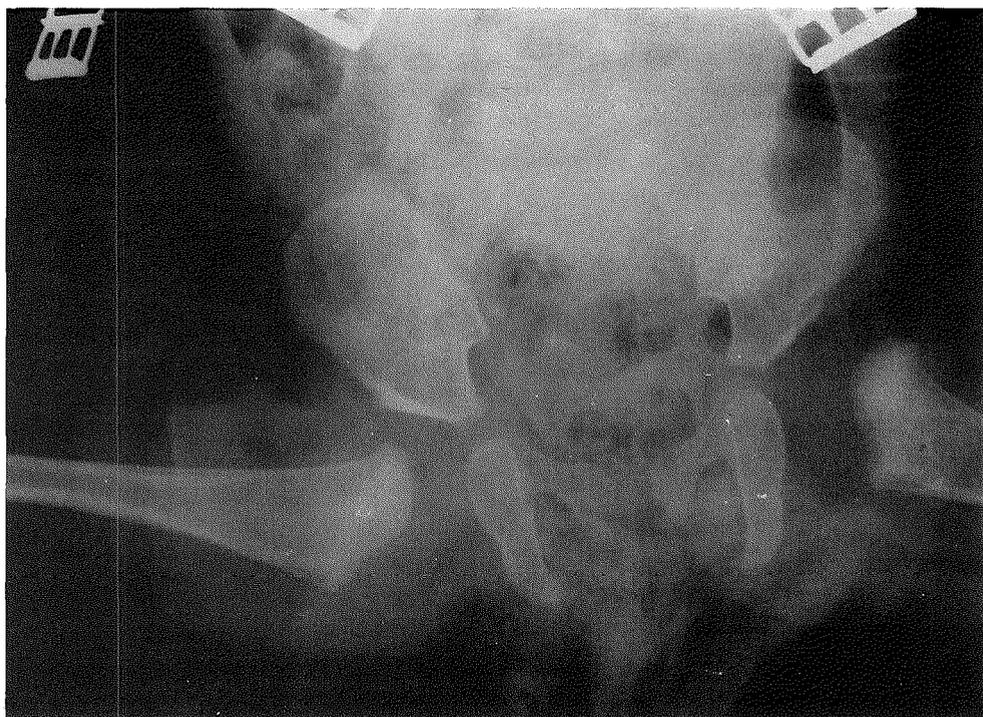


FIG. 14

Radiogram in the Pavlik harness. The left hip is still dislocated, but reduced in the harness within three weeks.

Positive Ortolani/Barlow tests at birth

Hips that showed positive Ortolani/Barlow tests at birth according to investigator I, were treated according to the protocol of our department. An a.p. pelvic radiogram was obtained, and they were re-examined clinically and by ultrasound after 1 and 3 weeks. If a positive Ortolani/Barlow test persisted beyond three weeks, treatment was instituted, in general in a Pavlik harness.

An X-ray was made in the harness to determine the position of the femoral head (Ramsey et al 1976) (Fig. 14), and the infant was checked clinically and by ultrasound at regular intervals until the hip had stabilised and an adequate development of the hip joint was obtained by ultrasound criteria. At that time an X-ray of the pelvis was taken. If this also showed a well-developed joint with sufficient coverage of the femoral head by the acetabulum and an acetabular index within normal limits, the child was gradually weaned from the abduction device.

Hips that had stabilised spontaneously were seen back at four months, when an X-ray was obtained.

Comparison between the radiogram and the ultrasound image at follow-up

The follow-up protocol offered the opportunity to compare pelvic radiograms at the age of at least six months with the ultrasonogram obtained at the same time.

For this part of the study, an additional group of twenty-five babies of at least six months old was included. These babies had been referred to our department for evaluation of their hips, a sonographic examination by the author was performed, and radiograms were obtained as well. These examinations were carried out in an identical manner to the protocol described previously.

Hips that were normal at birth in all respects

It was considered beyond the scope of this study to follow all infants that were normal at birth according to all criteria. This could have answered the question whether dysplasia can develop in a hip that is both stable and well formed at birth, but would have led to anxiety in more parents and a radiogram at a later age.

To determine the spontaneous development of the ultrasound appearance of the hip that is already well formed and stable at birth, 109 of these hips were reexamined by ultrasound after more than six months, however.

Statistical Analysis

In three instances, differing alpha values of groups of babies had to be compared. This was the case for left and right hips at birth, for boys and girls, and for positive versus negative family histories for CDH.

In these instances, Student's t test was employed to detect significance between the two groups of variables.

V.0 RESULTS

A total of 357 newborns was examined both clinically and by ultrasound on the same day by two independent examiners. There were 169 boys and 188 girls.

Examination was carried out routinely every day, so most infants were seen within 24 hours of birth.

In those cases where the examination was not carried out on day 0 or 1, this was usually due to the fact that the baby was born during the weekend, when no routine examination took place. Also, in a few cases, the child had to remain in an incubator for a few days due to general health problems.

In these instances the examination had to be postponed until the general health allowed it to take place.

The age at examination can be seen in Fig. 15.

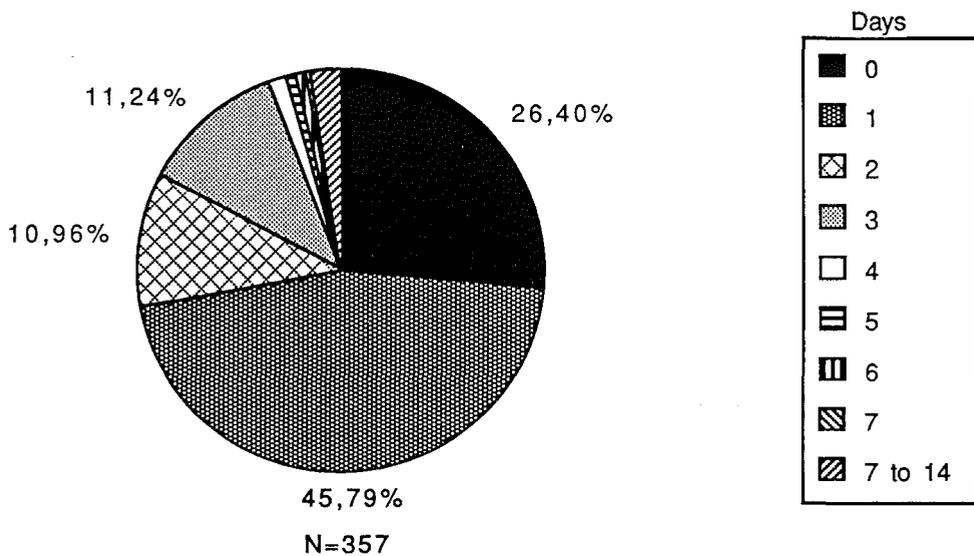


FIG. 15

Age at examination in days. 83% of the newborns were examined within the first three days of life.

Birth weight varied considerably from 1925 grams to 5010 grams.

The birth order of this group of newborns is shown in Fig. 16.

Twenty one children had a positive family history for congenital dysplasia of the hip, according to the mother. Usually this consisted of a second or third degree relative. In four cases first degree relatives were involved.

It appeared that most mothers were very well aware of the disorder they were asked about, especially if a near relative had been treated in a splint or operated upon in the first year of life.

Most babies were born in a vertex position, as can be seen in Fig. 17.

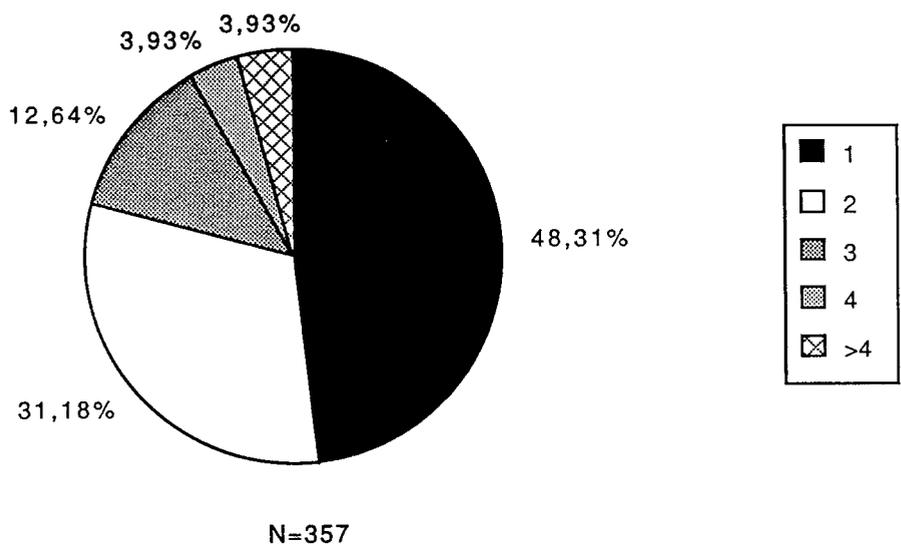


FIG. 16
Birth order of the total group of 357 newborns.

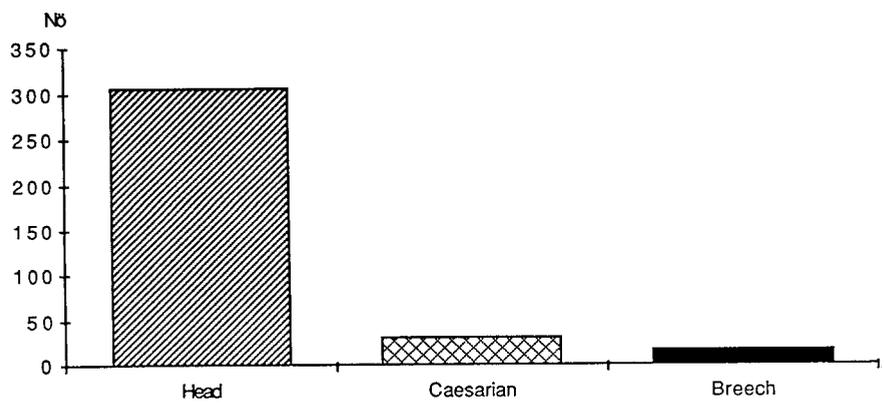


FIG. 17
Birth mechanism of the total group of 357 newborns.

V.1 Results of the neonatal clinical examination

Sixteen newborns, or 4.5% of the total screening population showed positive Ortolani/Barlow phenomena at birth, according to investigator I. Six of these were boys, there were ten girls.

In seven cases both hips were or could be dislocated, thus twenty-three hips (or 3.2% of all hips) showed a positive Ortolani or Barlow manoeuvre at birth. The birth order of these newborns can be seen in Fig. 18. Three had a positive family history for CDH.

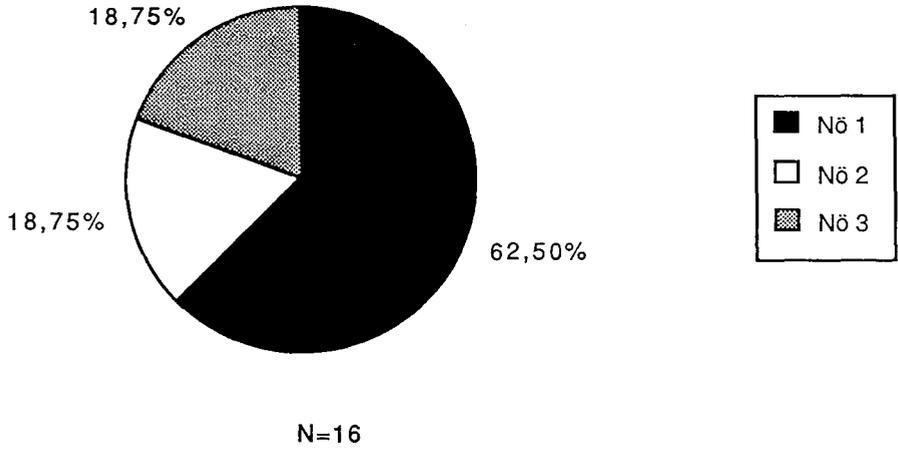


FIG. 18
Birth order of the sixteen newborns with dislocated or dislocatable hips at birth.

The birth mechanism of these newborns with positive Ortolani/Barlow signs showed a predominance of breech presentations (Fig. 19).

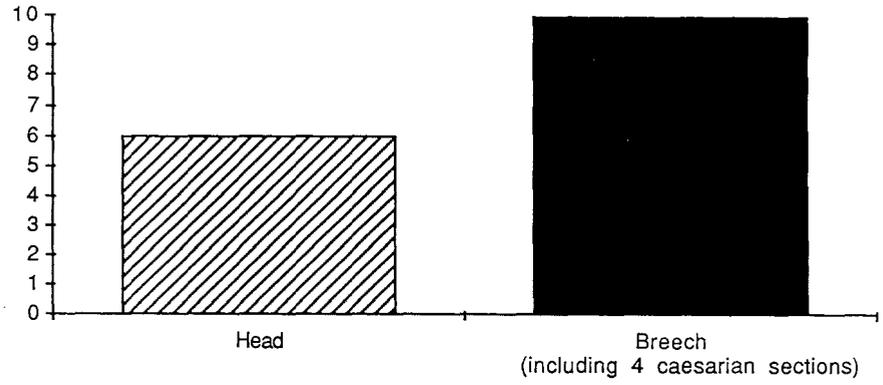


FIG. 19
Birth mechanism of babies with dislocated or dislocatable hips at birth.

Not all of these twenty-three hips were classified as abnormal based on the ultrasound examination, however. The overall impression was definitely good, with a well developed acetabular cavity in nine of these hips. The bony acetabular edge was slightly rounded in all of them, but this is a fairly normal finding in the newborn (Graf 1986). These nine hips had a well developed cartilaginous acetabulum as well. In four other hips the acetabulum looked slightly steep, these would be classified as type IIa according to Graf (1986). The distribution of the different angle measurements for all clinically abnormal hips with positive Ortolani/Barlow tests can be seen in the Fig. 20, 21, 22.

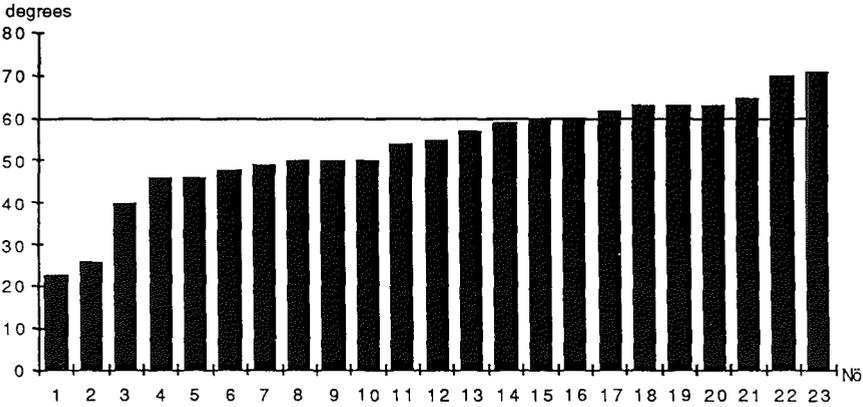


FIG. 20
Alpha angles of hips that were dislocated or dislocatable at birth. Although very low alpha angles were found in some hips, indicating a very steep acetabulum, cases 16-23 had a well-formed acetabulum on the ultrasound image.

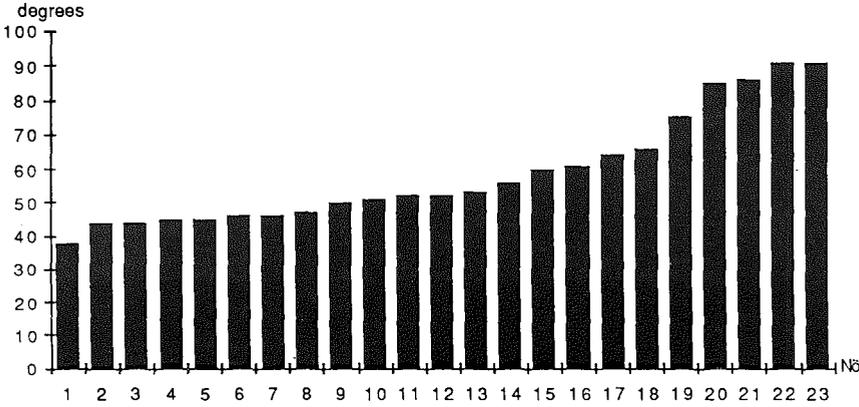


FIG. 21
Beta angles of hips that were dislocated or dislocatable at birth. The high beta angles correspond to hips that were dislocated at the time of the examination.

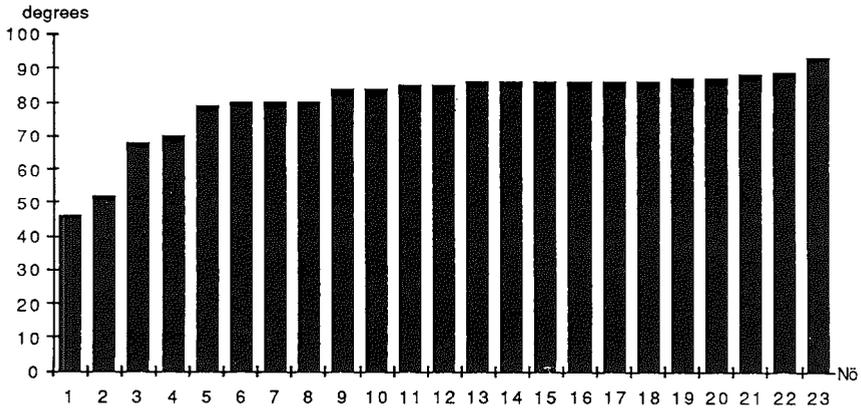


FIG. 22
Gamma angles of hips that were dislocated or dislocatable at birth.

During ultrasound examination in real-time, eighteen hips could be confirmed to dislocate out of the acetabulum. Five hips were considered stable under dynamic ultrasound examination, however. These five hips were stable according to the clinical examination after one week, and were not treated, but were followed at regular intervals. All five have developed normally over the course of their first ten months of live (Fig. 23).



FIG. 23
Normal hip at the age of 12 months. The infant showed a positive Ortolani test at birth, that was not confirmed by ultrasound. No treatment was instituted, since clinical examination was normal at one week.

The remaining eighteen that were confirmed to be dislocated or dislocatable according to ultrasound criteria, all remained so and required treatment in a Pavlik harness. Treatment was successful in obtaining and maintaining reduction in all cases. The duration of treatment nor its outcome could be related to the ultrasound image directly after birth. One other newborn had a severe adduction contracture at birth of the left hip. This child was later shown to have multiple congenital malformations, among which a teratological dislocation of that hip. The hip could not be reduced into the acetabulum manually, nor by overhead traction. Ultrasound as well as a pelvic X-ray showed a high dislocation (Fig. 24, 25). She was excluded from the study.

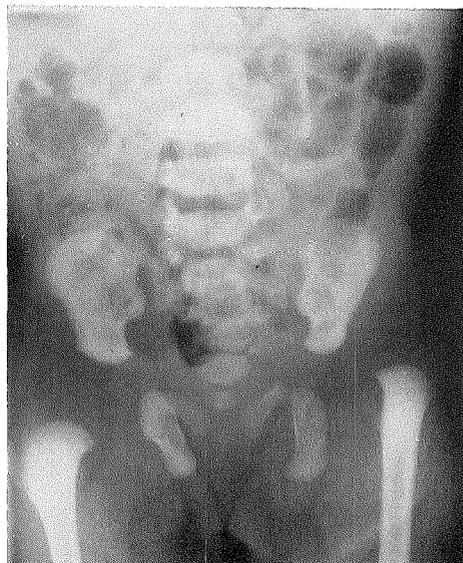
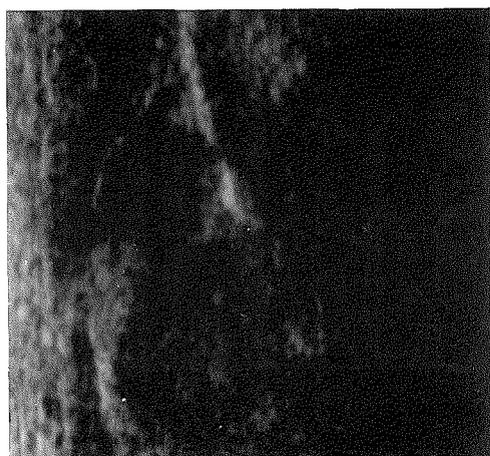


FIG. 24
Ultrasound image of a high dislocation (teratologic) of the left hip at birth. The femoral head abuts against the lateral pelvic wall.

FIG. 25
Radiogram of the same patient as in FIG. 24. This patient was excluded from follow-up.

V.2 Results of the neonatal ultrasound examination

For the total group of 357 newborns, the alpha angles for the right hip varied between thirty-seven and eighty degrees (average 63.9 degrees, standard deviation 6.8 degrees).

For the left hip, values varied between twenty-three and seventy-six degrees (average 62.5 degrees, standard deviation 6.9 degrees).

This difference is not significant according to Student's t test.

The average alpha angle for boys was 64.9 degrees (standard deviation 5.8), for girls 61.6 degrees (standard deviation 7.3).

According to Student's t test this difference is significant ($p < 0.005$).

Hips of newborns with a familiar predisposition for CDH had an average alpha angle of 59.9 degrees (standard deviation 10.1).

Newborns with a negative family history had an average alpha angle of 63.4 degrees (standard deviation 6.6). This difference is also significant ($p < 0.005$).

For hips with alpha values below sixty degrees, the average was 51.4 degrees (standard deviation 4.8), for hips with alpha values above and including sixty degrees, the average was 66.1 degrees (standard deviation 4.1).

All angles groupes together can be seen in Fig. 26.

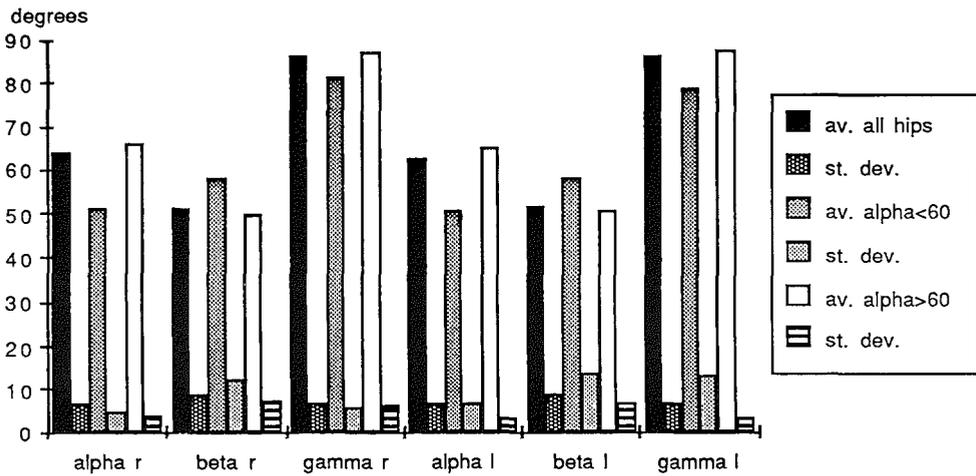


FIG. 26

Average angles with standard deviations of the total group of 357 newborns, of the group with an initial alpha angle < 60 , and the group with an initial alpha angle > 60 . Angles are given for the right hip (r) and for the left hip (l) separately.

V.3 Alpha angles below sixty degrees at birth

One hundred and seventeen hips (16.4%) in eighty newborns out of the total of 714 hips in 357 newborns had alpha angles less than sixty degrees at birth. In thirty-seven babies this occurred bilaterally, twenty-five had only the left hip affected, eighteen only the right hip.

Ten hips had an alpha angle of less than forty-four degrees (Graf type III or IV), thirty-two were between forty-four and fifty degrees (Type II dangerzone), seventy-five between fifty and sixty degrees (Type IIa).

The average alpha angles with standard deviation for this group of 117 hips can be seen in Table 1.

Table I

	Boys	Girls
Average alpha angle	55	54
Standard deviation	7	7

8 babies had a positive family history for congenital dysplasia of the hip. The first child was involved 34 times, the second 30 times, the third 10 times, the fourth 3 times, the seventh and eighth once each.

Isolated ultrasound abnormalities

Eleven newborns in this group of 117 hips had 15 hips that were found to be dislocated or dislocatable on clinical examination. The one newborn with the teratological dislocation was described above and excluded from the follow-up study.

Thus 101 hips remained, that were clinically without suspicion, in which only ultrasound had demonstrated a potential dysplasia.

Alpha angles varied for the right hip between 32-58 degrees (average 51, standard deviation 4.6), for the left hip they varied between 40-49 degrees (average 51, standard deviation 5.0) (Fig. 27).

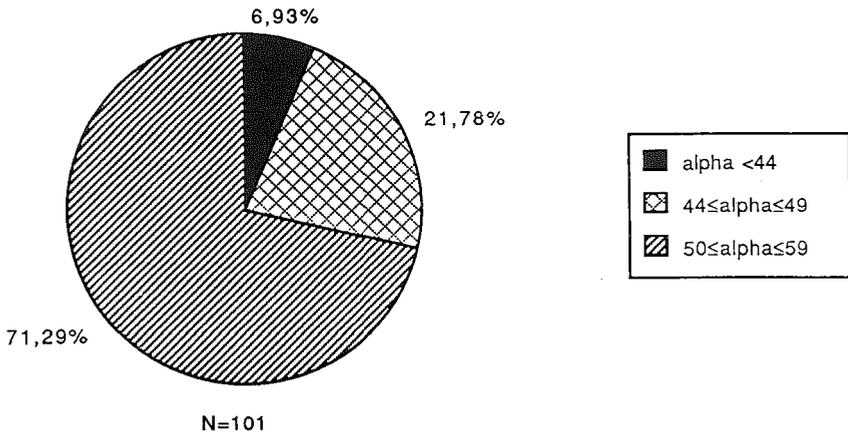


FIG. 27

Distribution of angles alpha <60 at birth, with a normal neonatal clinical examination.

As mentioned in the Patients and Methods section (chapter IV.0), these hips were not treated but reexamined after at least half a year.

At the end of the study, six children were not seen personally.

One had died shortly after birth due to a cardiac disorder, one was seen regularly by a pediatrician for unrelated reasons, the mother refused to have her child reexamined, one had moved to another area and was seen on my request at the age of ten months by an orthopaedic surgeon in that area. He could find no signs of dysplasia and considered an X-ray unnecessary. One was found out to have moved back to Pakistan with its parents, and two could not be traced at all.

Thus ninety-three hips (92% of the original group of 101) were reexamined personally. Follow-up ranged from 148-413 days, with an average of 241.5 days.

V.4 Ultrasound instability at birth

The term instability is often used in clinical examination, and often leads to confusion because it is ill defined.

What is meant in the context of this study, is the phenomenon of abnormal mobility of the hip joint under gentle pressure.

This ranges from an immediate dislocation upon the slightest pressure on the one end of the spectrum (Fig. 6), to less displacement of the femoral head with the labrum upon more (but gentle) pressure (Fig. 28 a and b).

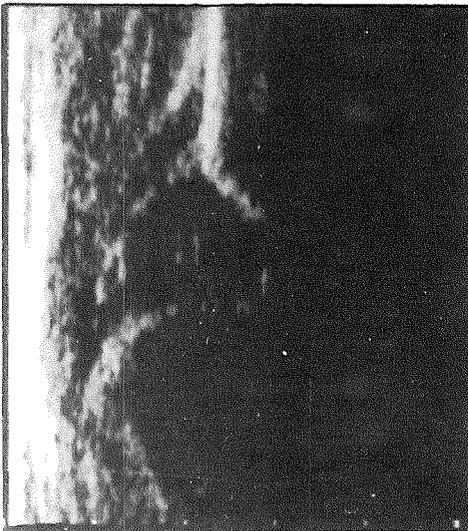


FIG. 28-A
Hip in the resting position.



FIG. 28-B
The same hip, now under gentle upward pressure. The femoral head is pushed up and laterally inside the acetabulum, with upward and lateral displacement of the labrum. Due to its dorsal and upward displacement, a steeper section of the acetabulum is seen.

Obviously, both the terms 'abnormal mobility' and 'gentle pressure' are subject to criticism, because they are not quantified. Any femoral head will move if enough pressure is applied.

In this study, both the ultrasound examination and the interpretation of the findings were done by the same investigator (investigator II, the author), who used the same technique in all children.

It therefore seems justified to discuss the findings of abnormal mobility of the femoral head as detected on the ultrasound at least for the purpose of this study, and draw a few modest conclusions since there were definite differences in mobility between hips.

In all ultrasound investigations in this study, the leg was gently moved through its range of motion. Subsequently, gentle upward pressure was applied to the leg during scanning in the frontal plane, and dorsal pressure during scanning in the transverse plane.

Instability was recorded if the femoral head was seen to slide up and down and/or back and forth inside the acetabulum with displacement of the labrum acetabulare (Fig. 28a and b).

The dislocation/reposition manoeuvre that occurred in eighteen hips with positive Ortolani/Barlow tests at clinical examination was seen beautifully in the dynamic investigation (Fig. 6). The femoral head dislocated in a dorso-cranial direction in all cases.

In all hips that exhibited this phenomenon, ultrasound showed the femoral head to be inside the acetabulum as long as the newborn was asleep. It slipped easily out of the joint as soon as the child got aroused and flexed the leg spontaneously. In these cases, ultrasound visualized and recorded an otherwise subjective, tactile sensation.

Thirty other newborns that were considered normal with stable hips according to clinical criteria, showed instability only on the ultrasonogram as defined above. Fourteen had this phenomenon bilaterally, nine on the left side and six on the right.

In all instances, the femoral head could be displaced in a craniodorsal direction, no hip manifested ventral instability.

There were eight boys and twenty-two girls, born at an average gestational age of 39.7 weeks (standard deviation 1.1 weeks).

Their average age at examination was 1.2 days.

Birth weight averaged 3392 grams (518.9 grams).

Three children had a positive family history for CDH.

Birth order: nine were first born, sixteen second born, two third born, and one was eighth in line.

Birth mechanism: twenty-seven were born head first, one was born by the breech, another required a caesarian section because of a breech position that could not be delivered naturally.

The alpha angles for the right hip ranged from 37-72 degrees, with an average of 57 degrees, for the left hip from 42-73 degrees, average 56 degrees.

All infants that showed isolated ultrasound instability (despite a normal clinical exam) were seen back at an average age of 240 days, no treatment had been instituted. **None** had any residual instability as defined above on the ultrasound at that age. All forty-three hips were absolutely normal at follow-up according to ultrasound, clinical and radiological criteria.

Thus this appears to be an innocent finding that is more prevalent in girls, and does not lead to a dysplastic development of the hip later in life.

V.5 Development of the bony acetabulum in the course of follow-up without treatment in hips that were clinically normal but showed alpha angles below sixty degrees at birth.

Bony cover of the femoral head by the acetabulum is expressed by the alpha angle as described by Graf (Fig. 11).

The development of the alpha angles of sonographically abnormal, but clinically normal hips at birth in the course of the follow-up without treatment can be seen in Fig. 29.

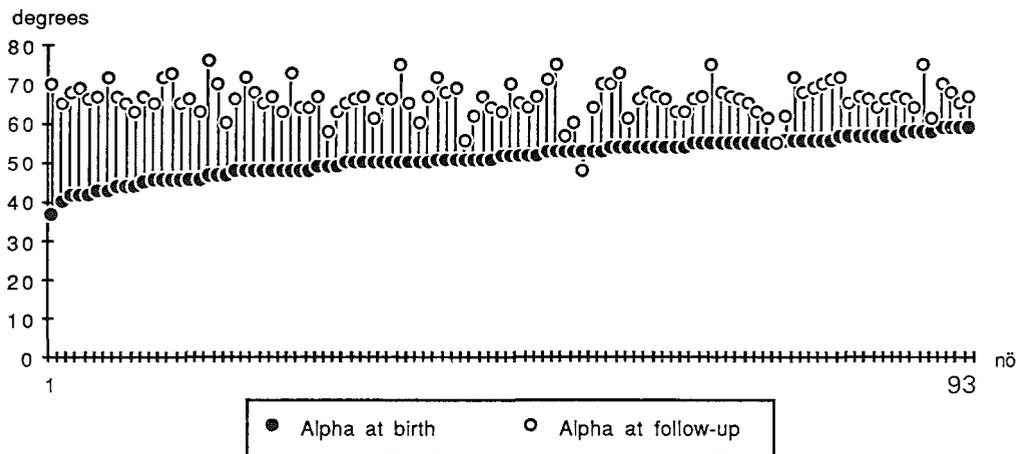


FIG. 29 Development of angle alpha in the course of follow-up without treatment in hips that showed alpha angles below sixty degrees, but were clinically unremarkable at birth.

As can be seen in the figure, angle alpha has spontaneously increased substantially in the course of follow-up in the majority of cases, indicating the increase in the ossification of the acetabular roof that has occurred.

V.6 Development of the cartilaginous acetabulum in the course of follow-up without treatment in hips that were clinically normal but showed alpha angles below sixty degrees at birth

One of the major advantages of ultrasound over other investigations is its ability to vi-

sualize the cartilaginous structures without the need for a contrast medium. Of all the other techniques that were described in the introduction, only Magnetic Resonance Imaging has the same properties, but its drawbacks were discussed. The cartilaginous acetabulum provides an important addition to the total cover of the femoral head at birth, and gradually ossifies in the course of development. Cartilaginous cover is expressed by the angles β and γ (Fig. 11, Fig. 13).

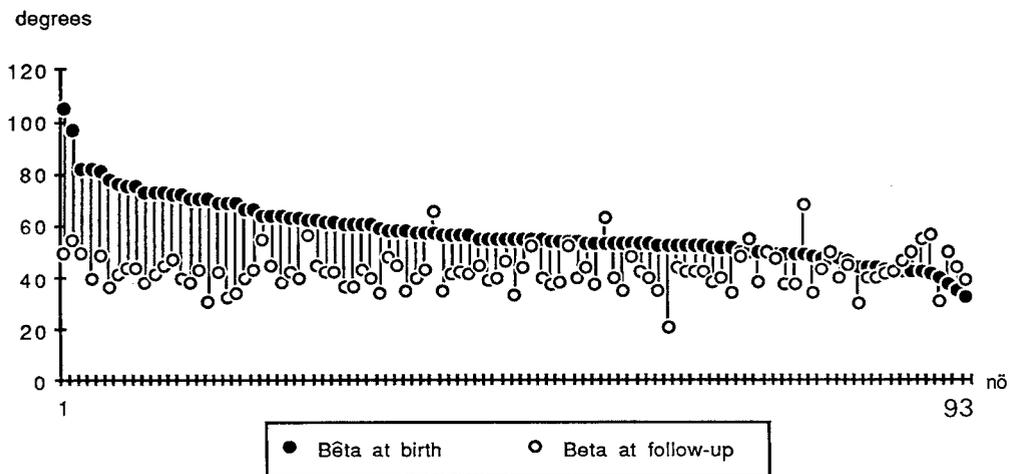


FIG. 30
Development of angle β in the same hips as in FIG. 29.

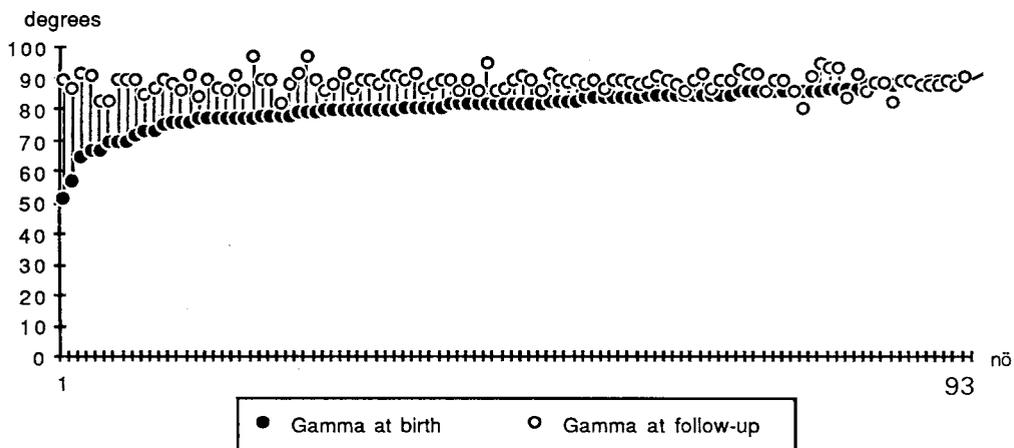


FIG. 31
Development of angle γ in the same hips as in FIGS 29 and 30.

The development of these angles indicates the changes that take place in the more deformable part of the acetabulum. Hips that subluxate or dislocate will primarily cause a deformation of these softer structures before the shape of the bone becomes altered.

The development of the cartilaginous roof of hips that were clinically normal, but had alpha angles below sixty degrees on ultrasound at birth can be seen in Fig. 30, 31.

Angle beta has decreased spontaneously in most cases, indicating a shift in favor of the bony cover, whereas angle gamma has remained more constant. This indicates that already at birth angle gamma provides relevant information about the final shape of the joint.

V.7 Development of ultrasound angles in hips that were normal at birth according to all criteria

Not all hips that were normal according to clinical **and** ultrasound criteria at birth were included in the follow-up study. A total of fifty-seven right and fifty-two left hips that were normal according to all parameters at birth was reexamined at an average age of 224 days, however.

Clinical examination showed no abnormalities at this age.

The development of the average ultrasound angles can be seen grafically in Fig. 32. In all hips that were reexamined, the alpha angle had increased, although only a few degrees. This indicates a consistent improvement of the bony cover of the hip. Angle beta had decreased slightly, indicating a shift in the acetabular roof in favor of the osseous development. No hip showed worse ultrasound angles at follow-up than at birth in this group.

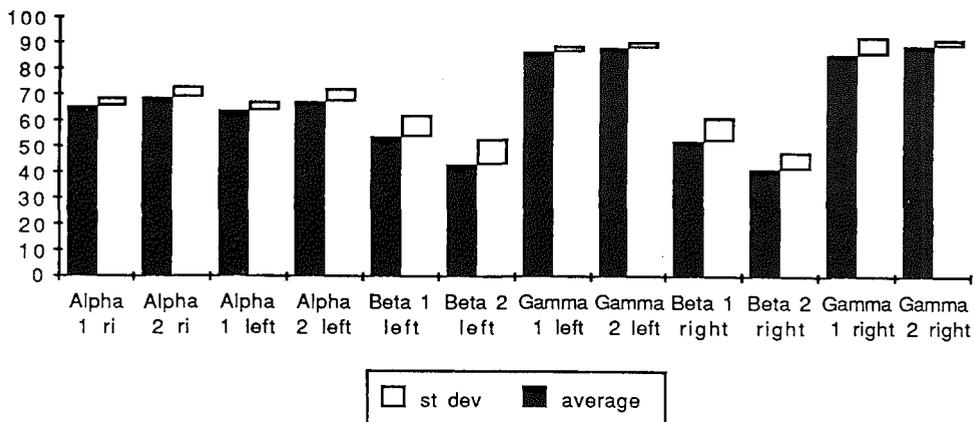


FIG. 32

Development of various ultrasound angles in hips that were clinically and sonographically normal at birth. 1 = at birth; 2 = at follow-up.

V.8 Radiological follow-up of clinically normal hips with alpha angles below sixty degrees at birth

At an average age at follow-up of 241.5 days an antero-posterior radiogram of the pelvis of these infants, that showed isolated 'ultrasound dysplasia' (including immature hips, type IIa) at birth was obtained. None had been treated.

Four hips had either a subluxation or definite dysplasia with an acetabular index of at least two standard deviations above the mean for age (Tönnis and Brunken 1968), with hypoplasia of the involved capital femoral epiphysis (Fig. 33, 34, 35) at follow-up, two right and two left. Two other hips showed milder dysplasia with an acetabular index between one and two standard deviations above the mean for age. Five other hips were judged to be 'borderline' as far as their development was concerned, with an acetabular index within one standard deviation above the mean value for age (Fig. 36). The remain-



FIG. 33
Dysplastic right hip at follow-up. The child had been considered clinically normal at birth, but had an alpha angle of the right hip below sixty degrees.



FIG. 34
Dysplastic left hip at follow-up, despite a normal neonatal clinical examination.



FIG. 35
Bilateral dysplasia at follow-up.



FIG. 36
Borderline development, with acetabular indices within one standard deviation above the mean for age bilaterally.

ning eighty-two hips were radiologically, clinically and sonographically normal at follow-up (Fig. 37).

The distribution of the various acetabular indices of the left and right hip at follow-up separately can be seen in Fig. 38 and 39.

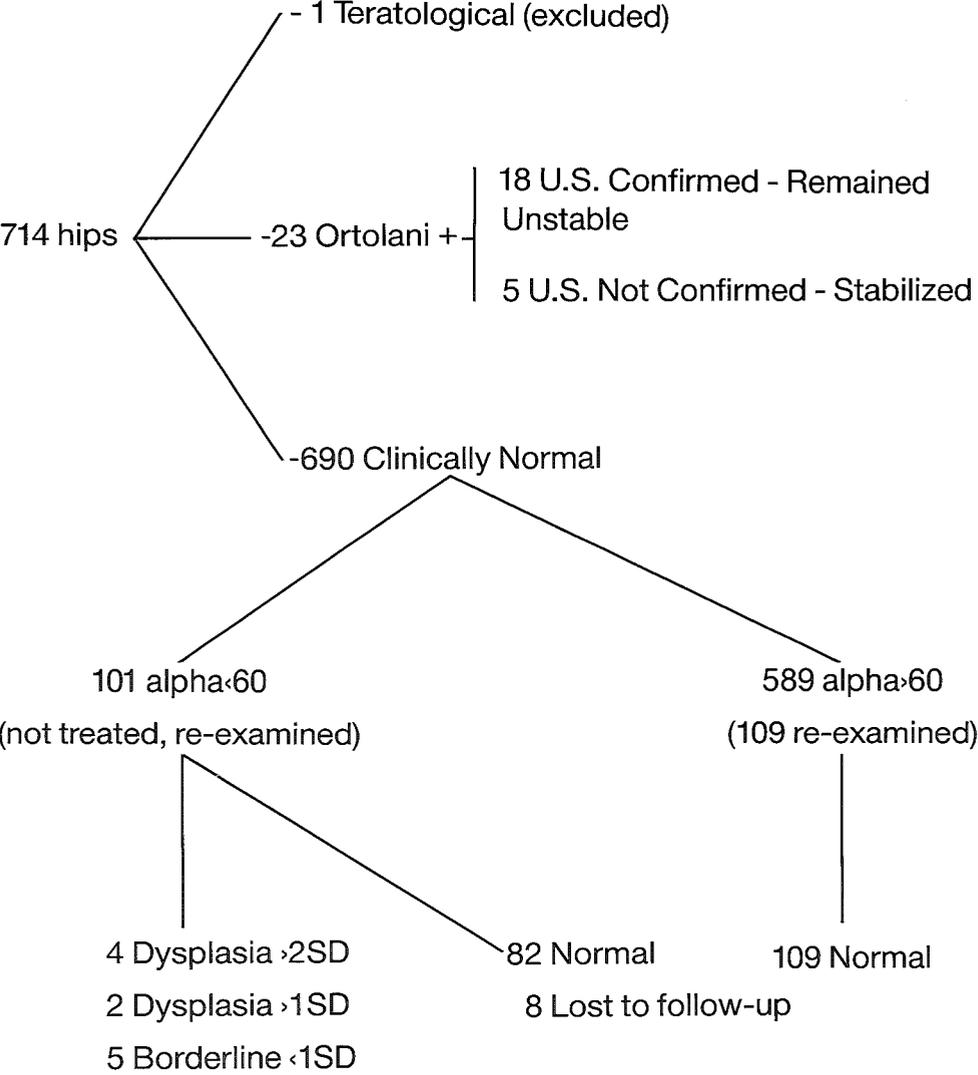


FIG. 37
The whole group of 357 newborns, and the findings at follow-up.

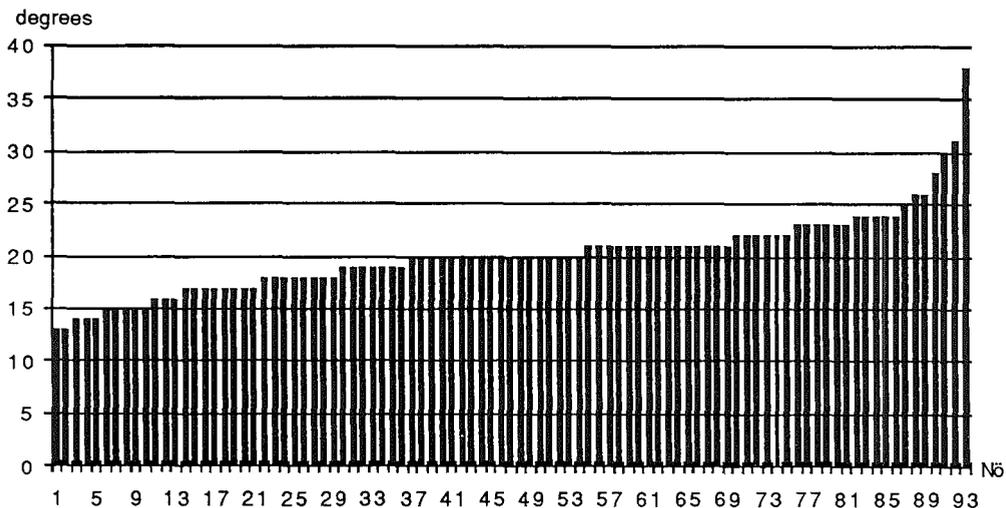


FIG. 38
The distribution of the acetabular indices for the right hip at follow-up. These were all hips that had a normal neonatal clinical examination, but showed steepness of the acetabulum ($\alpha:60$) on the neonatal ultrasound.

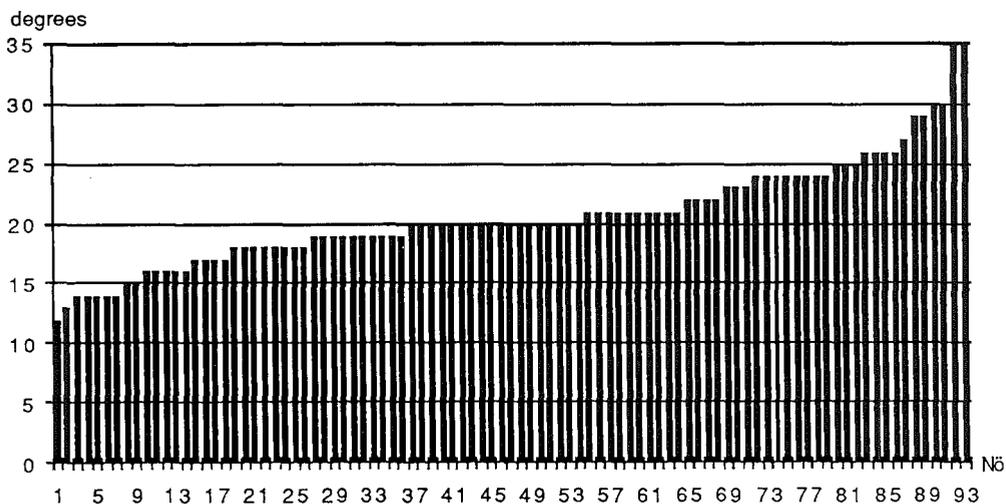


FIG. 39
The distribution of the acetabular indices for the left hip at follow-up. These were all hips that had a normal neonatal clinical examination, but showed steepness of the acetabulum ($\alpha:60$) on the neonatal ultrasound.

Comparison between the eighty-two normal hips and the four most dysplastic hips at follow-up

The four hips that showed the most evident radiological dysplasia at follow-up, with an acetabular index that was at least two standard deviations above the mean for age, were

studied specifically. Two of these children were born by caesarian section (both for breech presentation), the other had a bilateral dysplasia and was born by vertex presentation. Their acetabular indices at follow-up were 38, 35, 35 and 31 degrees. There were two right and two left hips, all of these patients were girls.

One of them was first born, two were second children. One child had a familiar predisposition. They were born in February, May and July. Birth weight varied between 2970 and 4380 grams, gestational age between 38-40 weeks. One child showed a disorder of its lower extremity at birth, a pes calcaneus. All other clinical parameters were normal at birth. The alpha angle at birth of the involved hips was 48, 53, 55 and 55 degrees.

The alpha angle at birth in the eighty-two hips that normalized varied between 37-59 degrees. Six children in this latter group showed a familiar predisposition for congenital dysplasia of the hip, nine children were born by a caesarian section. No disorders of the lower extremities were found at birth in this group.

Out of all children with an unremarkable clinical examination at birth, with no familiar predisposition, that were born by the vertex, that **had** an alpha angle below sixty degrees at birth, only one had a true dysplasia at follow-up.

No remarkable difference between the two groups could be found in alpha, bêta, or gamma angles.

V.9 Relationship between the radiological and sonographical criteria for hip development at follow-up

At follow-up the relationship between the ultrasound measurements of the bony acetabulum (alpha angle) and the acetabular index was assessed as can be seen in Fig. 40 and 41.

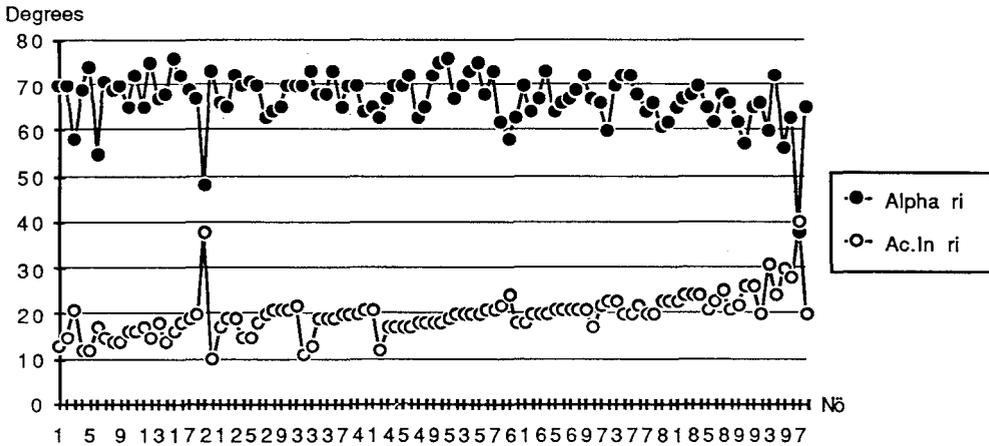


FIG. 40 Relationship between the alpha angle and the acetabular index for the left hip at follow-up.

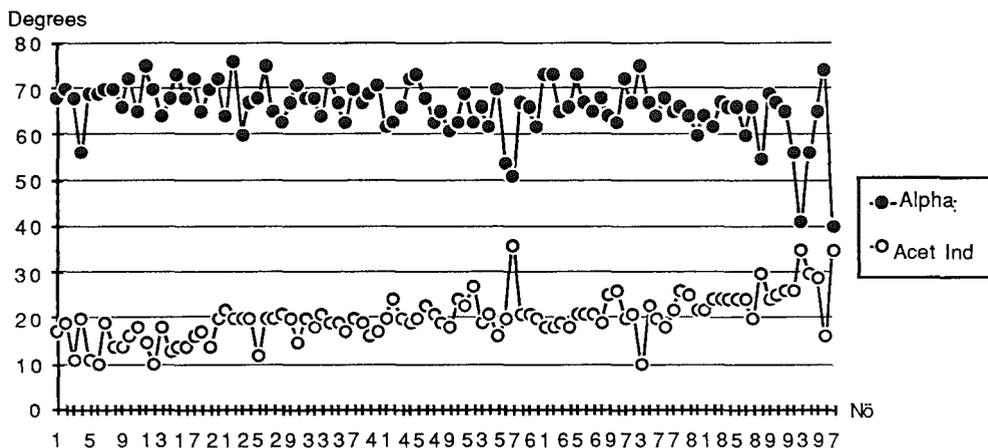


FIG. 41
Relationship between the alpha angle and the acetabular index for the right hip at follow-up.

	A	B	C	D	E	F	G	H	I	J	K	L
1	A.I. ri	A ri	A.I. le	A le	m/f	Age (days)		Xri	U.S.ri		Xle	U.S.le
2	26	55	25	58	f	237		D>1SD	D		B<1SD	B
3	26	57	26	56	f	368		D>1SD	D		B<1SD	D
4	20	66	25	63	m	222		N	N		B<1SD	N
5	40	38	35	40	f	184		D>2SD	D		D>2SD	D
6	31	56	30	56	f	377		D>2SD	D		D>1SD	D
7	28	63	35	41	f	246		D>1SD	N		D>2SD	D
8	31	60	30	65	f	153		D>1SD	N		D>1SD	N
9	27	65	32	45	f	155		N	N		D>1SD	D
10	24	72	30	55	f	191		N	N		D>1SD	D
11	25	65	26	65	f	229		B<1SD	N		N	N
12	24	68	25	67	f	378		B<1SD	N		N	N
13	50	35	24	66	f	184		D>2SD	D		N	N
14	38	48	16	74	f	189		D>2SD	D		N	N
15	20	70	20	54	m	152		N	N		N	D
16	24	58	20	66	f	186		N	B		N	N
17	21	58	10	75	f	335		N	B		N	N

TABLE II

This table shows the relevant findings at follow-up for sixteen children out of the total group of ninety-nine of whom a radiogram and ultrasound was obtained at the same time. A.I. ri = Acetabular Index of the right hip; A ri = Alpha angle of the right hip; A.I. le = Acetabular Index of the left hip; A le = Alpha angle of the left hip; m/f = sex; Age (days) = Age in days at follow-up; X ri = Status of the right hip based on the radiogram at follow-up; X le = Status of the left hip based on the radiogram at follow-up; U.S. le = Status of the left hip based on the ultrasound at follow-up; D = Dysplasia; B = Borderline; N = Normal.

There were seventy-four babies that were seen back according to the protocol of this investigation. Twenty-five others were included for this part of the study. These were children that had been referred to us because of suspicion of dysplasia, at an age of at least six months. Of these children an ultrasound examination was performed by the author exactly according to the principles outlined in the Patients and Methods section (chapter IV.0).

Thus radiograms and ultrasounds of ninety-nine children with an average age of 242.7 days were available for comparison.

According to radiological criteria, five hips showed borderline development as defined above. In these cases the ultrasound showed alpha angles of 58, 56, 63, 65 and 68 degrees (see Table II, case 2, 3, 4, 11 and 12).

According to ultrasound criteria, three hips showed borderline development with alpha angles of 58 degrees. Corresponding acetabulum indices were 25, 24 and 21 degrees (cases 2, 16, 17 in table II). Based on radiological criteria alone, fourteen hips would be classified as dysplastic, six with an acetabular index of at least two standard deviations above the mean for age, eight were between one and two standard deviations. One hundred and seventy-seven could be classified as normal.

Based on ultrasound criteria, thirteen hips would be classified as dysplastic (see Table II cases, 2, 3, 5, 6, 7, 9, 10, 13, 14, 15), hundred seventy-nine as normal (Table III).

The three hips that were normal according to ultrasound criteria, but dysplastic on the X-ray, as well as the one case in which the X-ray was normal with dysplasia of the ultrasound can be found in the table II (cases 7, 8 and 15).

Table III Comparison of radiogram vs. ultrasound per hip

	Radiogram Dysplastic	Radiogram Normal	
U.S. Dysplastic	11	1	12
U.S. Normal	3	176	179
	14	177	191

If we consider the radiogram at this age to be the most reliable parameter available to judge the development of the hip joint, a sensitivity for the ultrasound examination of 78.6% (11/14) can be calculated, a specificity of 99.4% (176/177), a positive predictive value of 91.7% (11/12) and a negative predictive value of 98.3% (176/179).

V.10 Clinical examination in hips that were dysplastic at follow-up

The age at follow-up of hips that were found to be dysplastic ranged from 153-368 days, with an average of 214.3 days. At follow-up examination, all clinical findings were re-

corded before the ultrasound was performed. Of three unilateral dysplasias, the examination showed a marked limitation of abduction in flexion.

In addition, one of these babies showed a rotatory malalignment of the involved leg, one also had a pes calcaneus.

The one child with the bilateral dysplasia at follow-up, plus one child with a unilateral dysplasia showed no remarkable findings at clinical examination.

Interestingly, all children had been seen regularly at the child care centers for periodic check-ups. In none of these instances any suspicion about the hips had been expressed by the examining doctors.

V.11 The ossific nucleus

The appearance of the ossific nucleus in the femoral head is an important parameter for skeletal development in conventional radiography. Graf (in Tönnis 1987) states that it appears two to three weeks earlier on the sonogram than on the radiogram.

At birth seven babies showed an inhomogeneous ossific nucleus on ultrasound on both sides. Three others had a unilateral ossification of the femoral head enough to be seen on ultrasound.

At follow-up all children showed an ossific nucleus on both sides, except for three at the respective ages of 92, 174 and 190 days. Four had only minimal ossification of the nucleus at the ages of 148, 163, 184 and 219 days. This study was not designed to determine the time of appearance of the ossific nucleus exactly, however.

In all cases of true dysplasia (verified by X-ray) there was a marked difference in the size of the ossific nucleus between the involved and the healthy side, the dysplastic side always being the smaller one.

VI.0 DISCUSSION

Congenital dysplasia of the hip remains a very crippling disorder if diagnosed and treated late.

Much effort has been invested into the earlier diagnosis and treatment, by means of carefully executed screening programs. Until now, screening for congenital dysplasia of the hip has focused on the detection of a dislocation or dislocatability of the femoral head predominantly by means of two clinical tests, Ortolani's (1937) and Barlow's (1962) manoeuvre.

This had been based on the -unproven- assumption that dysplasia, with or without dislocation of the hip in later life is a progression from a dislocation or dislocatability of the hip at the neonatal age.

The technique of the clinical examination requires skill and experience, and depends for a large part on whether the baby was at rest and relaxed at the time of the examination. Also, an examination that was positive at one time for the disorder may be negative only little later, even if it was carried out by the same investigator (Barlow 1962).

In general, clinical screening involves subjective, tactile sensations that are hard to share with others. Adequate stability as well as lack of it in the neonatal hip joint remains hard to standardise. A definite luxation, that can be reduced with the characteristic Ortolani 'clunk' will probably be recognized by the majority of experienced examiners, as well as the reduced hip that can be dislocated by means of the Barlow procedure.

Much harder to objectively record are the 'sliding hips' or milder losses of stability that for instance Tönnes (1987) describes. Also, the relevance of some of the physical findings is not well known.

Although Cunningham et al (1984) state with respect to newborn examination, that 'clicking and grating of the hip are important signs which require systematic follow-up', Sommer (1971) concludes that a 'dry hip click' resolves spontaneously without sequelae.

A number of clinical screening studies have claimed almost total success in preventing late-diagnosed cases, as well as almost uniform good results of early treatment (von Rosen 1956, 1962, Fredensborg 1976a,b, Palmén 1961, 1984, Miranda et al 1988, Hadlow 1988), although other reports have been far less encouraging, describing as many late detected cases as or more than before screening was started (Robertson 1984, Mackenzie 1972, Williamson 1972, Bjerkreim 1974).

A prevalent theory to explain late detected cases of dysplasia is that the shape of the acetabulum at birth plays an important role in the further development of the hip (Wynne-Davies 1970, Walker 1971, Cyvin 1977, Davies and Walker 1984). A steep acetabulum of a stable hip joint at birth could, in certain cases, lead to a later dysplastic development of the hip. According to Wynne-Davies (1970) these varying acetabular shapes are inherited as a multiple gene system. Obviously, these cases of 'congenital' dysplasia of the hip can never be detected by means of physical examination at birth, thus explaining at least a number of late diagnosed cases in spite of well executed screening programs.

There has long been a heartfelt need to visualize the infant hip joint in a non-invasive manner. Standard radiography is not considered very reliable at a very young age, probably until the age of \pm three (Krepler et al 1982) or four months (Owen 1968), although Putti (1933) advised its general use in the newborn.

Strict rules concerning rotation and tilt of the pelvis of the child have to be applied when a pelvic radiogram is taken of an infant, otherwise a considerable distortion of the projected image may occur (Tönnis and Brunken 1968). The technique of Andrén and von Rosen (1958) was never used extensively, probably due to the fact that it requires some force to keep the hip dislocated.

Before the age of three or four months, much information about the shape of the acetabulum can be obtained by means of arthrography. That this technique involves an invasive procedure that, certainly in the young child, has to be done under general anaesthesia, and also utilises ionising radiation is considered a major drawback for its use other than on strict indications.

Ultrasonography has provided a new development. It has emerged in the recent literature as a reliable technique to determine the shape of the hip joint in different planes, through a harmless and non-invasive investigation (see introduction to the subject, chapter II.10). Also, since the examination is done in real-time, it enables the investigator to move the hip through its range of motion and to test its stability. Subjective, tactile sensations ranging from definite dislocation to mild laxity can be objectively recorded and studied.

Dr. Graf, of Austria more than anyone else has developed the technique in pediatric hip disease, and has described its indications. His lateral approach to the hip, that provides an image in the frontal plane, and his detailed classification system, based on the appearance of the bony and cartilaginous acetabulum has been extensively followed. He advises the use of a 5 or 7.5 mHz (7.5 in newborns) linear array transducer placed parallel to the body axis on the region of the greater trochanter, with the infant in a positioning apparatus that holds it in the lateral position.

Although the region of the greater trochanter should be visualized, different degrees of flexion or extension of the legs hardly influence the image (Graf 1986, Zieger 1986). Correct imaging depends on finding the standard plane, in which the lateral pelvic wall projects as a straight line (Fig. 8, 9, 10). Failure to obtain this standard plane leads to a distortion of the projected image (Zieger 1986).

It appears from the literature, as well as from our own experience, that the upper age limit for this technique is between one year and eighteen months. After this age the ossific nucleus of the femoral head has become too big to allow clear visualization of the depth of the acetabulum. Standard radiography then provides more information than ultrasonography.

Although the global impression of the joint, including the dynamic examination is most important (Graf 1986, Zieger 1986, Zieger et al 1986), angles alpha and beta were introduced by Graf in addition for 'fine tuning' of the classification (Fig. 11). The alpha angle represents the most stable, least deformable cover of the femoral head. Zieger et al (1986) introduced angle delta, formed by the Baseline or Pelvic Wall Line and a line pa-

parallel to the osseous part of the acetabulum next to the acetabular rim, and the vector line, a line described to go parallel to the Baseline through the center of the femoral head. Morin et al (1985) described percentage of head coverage to determine acetabular development. These measurements do not seem to be an addition to Graf's system, and were not followed by other authors. Harcke et al (1984) and Boal and Schwenker (1985) use sector scanning and describe an additional transverse approach to the hip, with the transducer on the greater trochanter perpendicular to the body axis. This provides a more three dimensional image of the hip than the technique of Graf, which may be helpful in determining the position of the femoral head in cases of subluxation or dislocation, and was also employed in the present study, although with linear array. Their system does not offer such a differentiated interpretation of the shape of the acetabulum, however. Novick et al (1983), in addition to obtaining a longitudinal and transverse lateral image, place the transducer on the medial aspect of the thigh, with the leg in slight abduction. They use sector scanning as well. This approach has not been extensively followed by others. It was attempted by the author but seemed difficult to interpret for anatomical detail. There may be some use for this technique in children that are treated in casts or other abduction devices where only the region of the pubis is free. In general, the use of linear array rather than sector scanning equipment is advised since the images show more detail, especially of the superficial structures, and are easier to interpret (Graf 1986, Commission of the European Communities 1987).

Only Graf's system allows for fine differentiation of different hip types. All angle measurements lead to measurement errors, but for the alpha angle of bony inclination this error seems acceptable, especially if one investigator performs both the ultrasound examination and the measurements. This angle is defined by two lines, drawn between bony landmarks, that are relatively reproducible. The beta angle is determined by a less clear-cut cartilaginous landmark. Zieger (1986) found 4 ± 2 degrees variation in the alpha angle if images of the same hip were made by different ultrasonographers, but evaluated by the same reader. The influence of the reader was 6.5 ± 2 degrees for angle alpha. Angle beta showed variations of 6 ± 9 and 13 ± 5 degrees respectively. All measurements showed a high degree of consistency if the same reader re-evaluated the same image after an interval of two months.

According to Graf's criteria, the alpha angle has to be at least sixty degrees for a hip to be classified as definitely normal. Below the age of three months an alpha value between fifty and sixty degrees may still be acceptable, but these hips (type IIa, 'physiologically immature') should be followed at regular intervals, since rapid deterioration may occur (Graf 1986, van Moppes and de Jong 1988b, Tönnis 1989). Hips with alpha angles below fifty degrees at birth are thought to deteriorate without exception if they are not treated immediately (Graf 1986, Schuler and Rossak 1984, Tönnis 1989). Ultrasonography is considered to be 'more than twice as successful' in detecting pathological joints than clinical examination in the newborn, and has led to abduction treatment in twice as many cases as before neonatal ultrasound examination was started (Tönnis 1989). Also, Schuler (1987) states that, since the introduction of ultrasound screening, therapy could be started earlier in 80% of the cases. This had not led to an increase in

the overall number of infants requiring treatment, however. Although clinical screening of newborns is known to have its limitations, the danger exists that newborn hips will be judged on their ultrasound appearance exclusively. Ultrasound will only be useful as a screening instrument in newborns if it detects hips at the newborn age that will proceed to definite dysplasia, but cannot be diagnosed by clinical examination alone. Therefore, the natural history of untreated ultrasound abnormalities in hips of newborns **that are clinically normal** must be known. This problem is not addressed specifically in other studies on ultrasonography and has been the purpose of the present investigation.

Few other studies have dealt with the value of ultrasonography as a screening tool in an unselected group of newborns, although the Commission of the European Communities on Radiological Mass Screening within the member states advises against the use of ultrasound for general screening purposes on financial grounds (Kramer 1987a,b). The exact cost remains obscure, as well as the benefits, however.

Berman and Klenerman (1986) studied 1001 neonates clinically and by ultrasound for congenital dysplasia of the hip. All newborns were studied sonographically by one examiner (a radiologist) using 7.5 MHz sector scanning and clinically by the other (a pediatrician). Both referred to an orthopaedic surgeon if abnormalities were found. The orthopaedic surgeon was aware of the fact that abnormalities were found, but did not know the source of referral (whether from the ultrasonologist or from the clinician). Ultrasound showed only 2.8% of hips to have alpha values below sixty degrees. It could confirm the normal shape of the hip joint in a number of cases that had a positive Ortolani/Barlow test. These hips developed normally without treatment.

Eller and Katthagen (1987) suggest a general clinical and ultrasonographic screening program for all neonates in the first week of life in order to avoid late diagnosis and treatment. They do not provide data to support this suggestion, however, and are opposed by van Moppes et al (1988) on economic grounds.

Exner (1988) studied an unselected group of 615 newborns with ultrasound. He found 15.3% of a total of 1230 newborn hips to have alpha values below sixty degrees. Treatment criteria were based on the Graf measurements (1986) exclusively. Hips with alpha values between fifty and fifty-nine degrees (type IIa) were followed without treatment at regular intervals, and normalized in all forty instances that were reported. He proposes a cut-off value of fifty-five degrees instead of sixty for a hip to be classified as abnormal, in order to decrease the number of infants requiring follow-up. No data are provided about the natural history of these hips since all hips with alpha values below fifty degrees were treated regardless their clinical status.

Langer (1987) published a relatively large series of 1460 unselected newborns that were screened clinically and by ultrasound by two independent investigators. The examination was performed routinely between the third and tenth day after birth. Approximately 24% of hips had an alpha value below sixty degrees. All hips with alpha values below fifty degrees (type 2c, 3 and 4) were referred to the Orthopaedic Department for treatment, so the natural history of these hips cannot be determined based on this study. Benz-Bohm et al (1987) studied 162 unselected babies by ultrasound according to Graf's principles, mostly within the first two months of life (so this is not a newborn

screening program). The ultrasound investigation was performed without knowledge of the clinical or anamnestic data. They found alpha values between fifty and sixty degrees in 12% of the studied infants, only less than one percent had an alpha value below fifty degrees. In these latter children (only three) there were risk factors such as familiar dysplasia, a positive Ortolani sign or rotatory disturbances of the lower extremities. They conclude that ultrasonography is not a useful screening tool, but that children with established risk factors should be scanned.

Castelein and Sauter (1988), in a pilot study to this thesis, studied 307 unselected newborns. Both clinical and ultrasound examinations were done by the author of the present thesis. Treatment was based on clinical criteria exclusively, as in the present study. All clinically normal hips with alpha angles below sixty degrees were followed clinically and by ultrasound at six week intervals. They found that 13.4% of all hips had alpha values below sixty degrees at birth. Only 3.7% of these latter hips, that were normal on neonatal clinical examination, developed dysplasia in the course of follow-up, however. Since in this study ultrasound and clinical examination were performed by the same investigator, it was felt that the results of either part of the examination might have been influenced by knowledge of the other part.

So for the present study the protocol was revised. The clinical examination was done by another orthopaedic surgeon with experience in pediatric hip disorders (Investigator I), the ultrasound examination was carried out independently by the author (Investigator II) without knowledge of the results of the clinical examination.

Clinically normal hips with alpha angles below sixty degrees were not treated, but re-examined after at least six months.

A longer period of follow-up without intervention could be regarded as even more informative, but, regarding the potentially crippling disorder that this study deals with, seemed unethical. Ultrasound examination in the newborn was performed with the child on its side in its own bed. A positioning device as described by Graf (1986) has been tried, but it appeared that the newborn was much more at rest and comfortable in its own environment. The positioning apparatus was used in the older infants that were re-examined at follow-up.

The average age at follow-up was 241.5 days (range 148-413 days). There were 356 newborns in the study, 169 boys and 187 girls. Clinical and ultrasound examination were carried out on the same day, the age at examination can be seen in Fig. 15. Eighty-three percent of the examinations took place within the first three days of life. Although it is unlikely that the ultrasound image changes significantly during the first week of life, the importance of an early clinical examination was stressed by many authors a.o. von Rosen (1956), Fredensborg (1976a,b) and Barlow (1962).

The large majority of children was born by a vertex presentation (Fig. 17), so they were not in a specific risk group regarding their birth mechanism (Wilkinson 1963). Almost 50% of the examined infants was first born (Fig. 16). Wynne-Davies (1970) shows that the first born child is affected almost twice as often as subsequent children.

Routine clinical examination yielded twenty-three hips that were dislocated or dislocatable at birth, or 3.2% of all investigated hips. Although originally we had designed the

study to allow for a distinction between dislocated and dislocatable hips, it soon became clear to us that this was mainly an artificial distinction.

Based on chance, the femoral head could be inside or out of the joint depending on the position and state of activity the child was in. So analogous to Mackenzie and Wilson (1981) all hips that either were or could be dislocated were considered abnormal. Birth mechanism in these cases showed a predominance of breech presentations (Fig. 19).

This figure of 3.2% of hips is higher than what is usually reported, although wide variations exist between different series (0.3-2%). No definite explanation for this phenomenon can be given, although the size of the group and the fact that a clinical screening was performed may have an influence. The dynamic ultrasound examination confirmed the dislocation or dislocatability of these hips in **eighteen** cases, without knowledge of the clinical examination. **Five** hips that had a positive Ortolani or Barlow test according to investigator I (the clinical investigator) were considered absolutely stable upon ultrasound examination by the author. These five hips were also stable according to the clinical examination at one week, and were not treated but checked periodically. All five hips have developed normally so far, there is no sign that they will develop dysplasia later (Fig. 23).

The remaining eighteen hips that were also considered dislocated or dislocatable according to ultrasound criteria, all remained so by clinical and ultrasound criteria at follow-up and were treated successfully in a Pavlik bandage. Although Berman and Klenerman (1986) suggest that dislocated or dislocatable hips with a well developed acetabulum on the ultrasound have a better prognosis than the ones with a more abnormal acetabulum, no such correlation between the shape of the acetabulum and the duration of treatment could be found in this material. An attempt was made to define the position of the femoral head inside the Pavlik harness by ultrasound as described by Grissom et al (1988), but it was difficult to obtain adequate images in the frontal plane due to the flexed and abducted leg. Perhaps this is easier if a sector scanner is used. In this study, all positions inside the Pavlik harness were confirmed by one a.p. radiogram (Fig. 14). The five hips that were not treated would be called 'spontaneously stabilised' in some clinical studies, or treated with 'success' in others. It remains debatable if these hips were dislocated in the first place, or whether they represent cases of false-positive results of the clinical examination. Thus, the overall prevalence of truly dislocated or dislocatable hips in this study could very well be 2.5% instead of 3.2%.

All hips with a sonographic confirmation of the positive Ortolani/Barlow test could be scanned both in the dislocated or in the reduced position. Ultrasound demonstrated the dislocation-reposition manoeuvre beautifully (Fig. 6). It appeared that with the hip in the reduced position, the acetabulum looked almost normal in about half the cases (Fig. 20). In these hips, ligamentous laxity must have played an important role since the acetabulum looked adequately developed in the standard plane. However, in the dislocated position, the ultrasound transducer had to follow the femoral head to a more dorsal section through the hip joint. In this plane the acetabulum looked much steeper than in the more ventral cut. This confirms the findings of Serringe and Kharrat (1982), and of Ponseti (1978) who described a defect in the posterior part of the acetabulum in cases

of congenital dysplasia. In the transverse image, all hips were seen to dislocate posteriorly. This is an interesting finding, since even computed tomography has been used (Edelson et al 1984) to determine the direction of dislocation.

Also, it appeared that in the cases of a dislocation or dislocatability, the hip usually was reduced well inside the acetabulum when the child was relaxed. As soon as it became more active, and especially upon sudden active flexion of the leg, the hip was seen to dislocate spontaneously in a dorsal direction. It seemed as if the child performed his own Barlow manoeuvre, which could indeed be counteracted by gradual and gentle abduction of the leg. This points to the role of the iliopsoas muscle in the pathogenesis of congenital dysplasia of the hip. Although Sommerville (1962) and Salter (1968) have stressed the importance of avoiding forcible extension of the hip in newborns, as this might lead to dislocation, flexion without abduction apparently can also force the femoral head over a shallow acetabular edge posteriorly, and should be avoided.

'Ultrasound instability' without a positive Ortolani or Barlow test occurred relatively often, in forty-three hips. The problems associated with defining this phenomenon were discussed in chapter V.4. Ultrasound is very sensitive in detecting minor forms of mobility of the femoral head inside the acetabulum, as was also described by Graf (1986) and Clarke (1986).

Girls were more susceptible to this phenomenon than boys, in a ratio of 2.71:1. Although Graf (in Tönnis 1987) differentiates between 'elastic deflection' which may even occur in fully mature hips, and a 'sonographic instability sign' in severely deficient hips, no such distinction could be made in the present material. At follow-up, at an average age of 240 days, none of these hips showed a dysplasia according to clinical, ultrasound or radiological criteria. All femoral heads at this time were stable inside the acetabulum on ultrasound. This type of slightly increased mobility must be considered to be an innocent finding that occurs predominantly in girls.

For the total population of 357 newborns, the left hip showed a slightly lower (or worse) alpha angle than the right (62.5 vs. 64.0 degrees). Although this trend is noteworthy, the difference is not significant as determined by Student's t test. The predominance of left sided CDH (Dunn 1976) apparently cannot be explained on the basis of a generally more shallow left acetabulum at birth. This study has also shown a higher (or better) alpha angle for boys than for girls (64.9 vs. 61.6 degrees). This difference is statistically significant according to Student's t test ($p < 0.005$). The predominance of girls with CDH in the population (Bjerkreim 1974, Dunn 1976) may be related to a tendency of girls to have steeper acetabula than boys. This is confirmed by the tables of Tönnis and Brunken (1968), who found lower (or better) mean values for the acetabular index at different ages for boys than for girls. Also, a significant difference was found between alpha angles of newborns with or without a familiar predisposition, the former showing lower (or worse) alpha angles. Other screening studies that have used ultrasound do not state these findings specifically.

Seventy-nine otherwise healthy, clinically normal newborns had 116 hips with an alpha angle less than sixty degrees. Thirty-seven times this occurred bilaterally, twenty-four times only on the left side, eighteen times only on the right. This group consisted of

twenty-two boys and fifty-seven girls. Girls apparently have steeper hips more often than boys, a fact also reported by Tönns (1989).

Thus, in this unselected group of newborns, ultrasound abnormalities were found in **sixteen percent** of all examined hips. This extremely high percentage of supposedly abnormal, or potentially abnormal hips in a healthy population is astonishing, especially when one considers the reported incidence of established hip dysplasia in the community, somewhere between 0.3 and 2.0% (Mitchell 1972, Hierton and James 1968). Nevertheless, using ultrasound, Exner (1988), Benz-Bohm et al (1987), Langer (1987) and Schuler and Rossak (1984) find similarly high percentages. If we exclude fifteen hips of newborns that had a positive Ortolani/Barlow test at clinical examination, this leaves 101 hips that were clinically without any suspicion of abnormality, but showed a true or potential dysplasia that could only be detected by ultrasound. The majority of these hips (72) had alpha angles between fifty and sixty degrees, and would thus be called type IIa or physiologically immature according to Graf (1986), with the possibility of spontaneous improvement. Van Moppes and de Jong (1988b) report that approximately 22% of these hips ultimately required treatment. Tönns (1989) finds a deterioration of these hips in 9% of the cases. On the other hand, Exner (1988) found no deterioration in type IIa hips (he reports forty), and Langer (1987) reported 98% to heal uneventfully without treatment.

The remaining hips (29 hips) had alpha angles below fifty degrees (Fig. 27). These hips are in the definitely dysplastic group according to Graf, but showed no clinical signs or symptoms !

At an average age at follow-up of 241.5 days, all children with a neonatal alpha angle below sixty degrees despite a normal neonatal clinical examination were reexamined, and an a.p. pelvic radiogram was taken. None had been treated. A follow-up percentage of 92% was obtained. The radiogram taken at this age of all infants, showed dysplasia of the hip in six cases. Four of these had rather severe dysplasia, with an acetabular index of more than two standard deviations above the mean for age (Fig. 33, 34 and 35). In this category, the hip will generally become worse if untreated (Tönns and Brunken 1968). The two other hips had less severe dysplasia, with an acetabular index between one and two standard deviations above the mean. Five hips had an acetabular index less than one standard deviation above the mean for age. In the material of Tönns and Brunken, only 2.5% of the hips in this latter category became worse if untreated. The further natural history of these hips cannot be gathered from this study, since they were subsequently treated. So in the end, 6.5% of the hips that, at neonatal examination, showed alpha angles below sixty degrees despite a normal clinical examination, were dysplastic at follow-up and required treatment (Fig. 37). They definitely constitute a different category than the eighteen hips that were dislocated or dislocatable at birth. This confirms the theories of Wynne-Davies (1970), Walker (1971), Cyvin (1977) and Davies and Walker (1984), and demonstrates conclusively that there are indeed two types of congenital dysplasia of the hip. One group consists of hips that are or can be dislocated at birth due to ligamentous laxity, the other has hips that are stable at birth and therefore cannot be detected by clinical screening. The term 'missed diagnosis' does **not** ap-

ply to these hips. Ultrasound at birth does not solve this problem, however, since many more hips are detected than become dysplastic. Using a lower cut-off value than sixty degrees (Exner (1988) suggests fifty-five degrees) would decrease the number of hips that have to be followed. If fifty-five had been used as the lower limit of normal in this study, sixty-seven hips would have required follow-up (instead of 101). No hip with an alpha value at birth **above** fifty-five became dysplastic, but it cannot be predicted from the angle measurements which ones will deteriorate. The lowest alpha angle at birth of a hip that ended up dysplastic was 48 degrees, whereas the lowest alpha angle at birth of a hip that normalized was 37 degrees. All other hips that could not be distinguished at birth by means of ultrasonography from the ones that became dysplastic, were absolutely normal according to all criteria at follow-up.

Only one infant in the ultimately (at follow-up) dysplastic group did not have associated risk factors, such as familiar predisposition or breech presentation. In the six hips, that were dysplastic at follow-up, the clinical examination at this time showed a limitation of abduction or abnormalities in the lower extremities such as pes calcaneus or a rotatory malalignment in three cases. It is interesting to note that the child care centers where all infants had been seen regularly on a routine basis had never mentioned these abnormalities, possibly due to the fact that the parents had always told that their child had an appointment at an orthopaedic department for its hips. The one child with the bilateral dysplasia and one with a unilateral dysplasia showed no remarkable findings at clinical examination at follow-up at all.

Although radiograms of the pelvis taken at this age are generally considered to be reliable (Kreppler 1982, Owen 1968), Kreppler (1982) argued that the tables of Tönnis and Brunken (1968) give values for dysplasia that are too high. The ultimate classification of the hip did not depend exclusively on these measurements, however. Shape of the acetabulum, lateral sclerosis of its edge, lateroposition of the femoral ossific nucleus and hypoplasia of this nucleus were all taken into account.

The development of the ultrasound angles can be seen in Fig. 29, 30 and 31. Angle alpha generally has become higher with time, indicating an improvement in the bony cover of the hip. Not much is known about the rate of ossification of the cartilaginous acetabulum during the first year of life and its reflection on the alpha angle, although the acetabular index on the radiogram decreases (= improves) with age (Tönnis and Brunken 1968). Similarly, it is very important that different ultrasound values for different age groups are established. It is interesting to note that angle beta develops almost in an opposite manner to angle alpha, indicating that these two phenomena supplement each other. Angle gamma, representing the total cover of the acetabulum up to the labrum, also increases spontaneously, especially in hips that showed poor cover at birth (Fig. 31). In these hips, apparently a process of growth takes place, that leads to a deeper and more horizontal acetabular cavity, whereas in hips that showed better overall cover from the start, the process of maturation is more a shift from cartilage to bone.

It was considered beyond the scope of this study to follow all hips that were absolutely normal at birth in all respect, both clinically and sonographically. This could have answered the question if dysplasia could develop in hips that neither have ligamentous lax-

ity nor a shallow acetabulum, but it would also have required a radiogram at a later age of a supposedly normal child. Sonographic development was followed in a group of 109 of these normal hips over a period of more than six months, however (Fig. 32). The same development of the average angles could be noted as in the group that was sonographically abnormal at birth, with a consistent improvement of ultrasound angles.

At follow-up a comparison was made between the radiogram and the ultrasound both taken at the same time. When compared with the radiogram as the best standard at this age, a sensitivity of the ultrasound examination of 78.6% was obtained, a specificity of 99.4%, a positive predictive value of 91.7% and a negative predictive value of 98.3%.

The relatively poor sensitivity is explained by the fact that the number of dysplastic hips is small, and that the three hips that were considered normal on the ultrasound, but dysplastic on the radiogram had an acetabular index of between one and two standard deviations above the mean. Although the alpha angles of these hips were above sixty degrees (table II), and they were therefore not classified as dysplastic, their bony acetabular edges were rounded, and the cartilaginous acetabulum was broad in two. This emphasizes the importance of judging the overall impression of the ultrasound image, which is true for radiograms as well. For the purpose of scientific study, angle measurements are necessary for objective recording however. Van Moppes and de Jong (1988a) calculate a sensitivity of 92.2%, but their abnormal group was much larger, since they performed their investigations on indication. Zieger (1986) in a detailed study including 271 abnormal hips claims a sensitivity of 100% for dislocated hips and a specificity of 98%.

Based on all findings combined, it must be concluded that ultrasonography at the newborn age should not be performed as a neonatal screening procedure in the general population, but that it should be reserved for hips with positive Ortolani/Barlow manoeuvres, and for close surveillance of infants with risk factors such as breech delivery and a positive family history. The questions asked at the beginning of this study (Chapter II.1) can be answered as follows:

1. Hips that are clinically normal at birth and show no risk factors, but have alpha angles below sixty degrees, almost never develop dysplasia.
2. Clinically normal hips, that show mobility of the femoral head inside the acetabulum on the ultrasound at birth, develop into normal hips.
3. In this study, at the age of six months, the sensitivity and specificity of ultrasonography as related to a pelvic radiogram was 78.6% and 99.4% respectively.

VII.0 SUMMARY AND CONCLUSIONS

In the present investigation the additional value of ultrasound screening of newborns for congenital dysplasia of the hip, over routine clinical examination was studied. The main question that has led to this prospective study was, whether neonatal ultrasonography could detect cases of hip dysplasia that would be missed by clinical examination of the newborn alone. In order to answer this question, an unselected group of 357 newborns was examined clinically and by ultrasound by two independent investigators. In this group, 116 hips showed an abnormal, or potentially abnormal ultrasound examination (Graf type IIa, III or IV). Fifteen of these hips had a positive Ortolani/Barlow sign at birth, so 101 hips with an unremarkable neonatal clinical examination had ultrasound abnormalities. These infants were not treated, but reexamined clinically, by ultrasound and pelvic radiography at an age of at least six months.

Six of these hips showed definite, established dysplasia at follow-up. Five other hips had an acetabular index that was within one standard deviation above the mean for age (Tönnis and Brunken 1968), and were termed 'borderline'. Eighty-two were normal according to all criteria at follow-up. Eight hips were not seen back personally. Three infants, with the four severest dysplasias at follow-up (acetabular index $>2SD$ above the mean for age) were compared for their ultrasound image at birth with the eighty-two hips that were normal at follow-up. No differences in ultrasound appearance of these hips could be found right after birth. Alpha angles at birth of the hips that became dysplastic were 48, 53, 55 and 55 degrees. The lowest alpha angle in the group of hips that became normal was 37 degrees. All dysplasias at follow-up occurred in girls, all but one showed risk factors such as delivery by caesarian section, breech presentation or familiar predisposition. Ultrasound could objectively record otherwise subjective findings as a positive Ortolani or Barlow manoeuvre accurately. In this study, twenty-three hips were thought to be either dislocated or dislocatable at birth according to the clinical investigator.

Five of these were stable according to the neonatal ultrasound examination, however. These hips were considered stable by both clinical and ultrasound standards after one week, and did not require treatment. They have developed normally over the course of their first ten months of life, with a normal clinical examination, normal ultrasound images as well as normal radiograms. The hips that were confirmed to be dislocated or dislocatable according to ultrasound at birth (eighteen hips), remained so after one and three weeks and required treatment.

Possibly the hips that were stable on the neonatal ultrasound in this study represent false-positive results of the neonatal clinical examination. Ultrasound could aid in the decision not to treat these essentially normal hips.

Ultrasound instability at birth (defined in Chapter V.4) in clinically normal hips occurred in eight newborn boys and twenty-two girls, in forty-three hips. Their alpha angles ranged from 37-73 degrees. These infants were seen back at an average age of 240 days, they had not been treated. None showed residual instability on the ultrasound at that time, all hips had developed normally according to clinical, ultrasound and radiological criteria.

At follow-up, a comparison was made between the radiogram and the ultrasound image obtained at the same time. For this part of the study, twenty-five other infants that were referred for possible hip dysplasia were included. Three hips had normal alpha values, but were dysplastic on the radiogram. These had an acetabular index between one and two standard deviation above the mean. In two of these, the overall ultrasound image showed signs of dysplasia such as rounding of the bony acetabular edge, and broadening of the cartilage. The X-ray confirmed ultrasound dysplasia in eleven cases. If the radiogram at this age is considered to be the ultimate standard, the sensitivity of ultrasonography in this study was 78.6%, the specificity 99.4%, the positive predictive value 91.7% and the neative predictive value 98.3%.

Conclusions:

- Ultrasonography should not be performed as a general neonatal screening procedure for hip dysplasia in clinically normal newborns that have no associated risk factors. In these children, although a wide variety of hip types with different acclivity of the acetabulum exists at birth, almost all hips will become normal with time.
- Ultrasound instability at birth in clinically normal hips is an innocent finding that does not lead to dysplasia in later life. It occurs more often in girls than in boys.
- Ultrasound is valuable in confirming a positive Ortolani or Barlow test.
- Newborns with associated risk factors such as breech delivery, caesarian section, or familiar predisposition, deserve to be followed at regular intervals by clinical examination and ultrasound, since they may develop dysplasia in the course of their first few months of life, despite a normal neonatal clinical examination.
- When compared to a pelvic radiogram at the age of six months, ultrasound shows a good sensitivity and excellent specificity, especially when the overall ultrasound image (and not just angle measurement) is taken into account.

VIII.0 SAMENVATTING EN CONCLUSIES

De aanvullende waarde van een echografische screening van pasgeborenen op congenitale heupdysplasie, ten opzichte van het routine klinische onderzoek, is onderzocht in de onderhavige studie. Kan heupechografie bij de pasgeborene, gevallen van congenitale heupdysplasie onderkennen, die worden gemist bij het neonatale klinische onderzoek? Om deze vraag te beantwoorden is een ongeselecteerde groep van 357 pasgeborenen klinisch en echografisch onderzocht door twee onafhankelijke onderzoekers. In deze groep toonden 116 heupen een afwijkend, of potentieel afwijkend echografisch beeld (Graf type IIa, III of IV).

Bij vijftien van deze heupen was het neonatale klinische onderzoek afwijkend, met een positieve handgreep van Ortolani/Barlow, dus 101 heupen toonden echografische afwijkingen ondanks een normaal klinisch onderzoek. Deze kinderen zijn niet behandeld, maar na ten minste zes maanden klinisch, echografisch en röntgenologisch opnieuw onderzocht. Bij het na-onderzoek bleken zes heupen een duidelijke dysplasie te hebben. Vijf andere heupen hadden een acetabulum index binnen één standaard deviatie boven de gemiddelde waarde voor deze leeftijd (Tönnis en Brunken 1968). Tweeëntwintig heupen waren normaal volgens alle criteria bij het na-onderzoek. Acht heupen zijn niet persoonlijk gecontroleerd. Van drie zuigelingen, met de vier ernstigste dysplasieën bij het na-onderzoek, zijn de neonatale echografische gegevens vergeleken met die van de tweeëntwintig normale heupen bij het na-onderzoek. Er kon geen verschil worden gevonden in het echografische beeld direct na de geboorte. De neonatale alpha hoeken van de heupen die dysplastisch werden bedroegen 48, 53, 55 en 55 graden. De laagste alpha hoek in de groep die normaal was bij het na-onderzoek, bedroeg 37 graden bij de geboorte. Alle bewezen dysplasieën bij het na-onderzoek traden op bij meisjes. Behoudens één bestonden er bij allen risico factoren zoals een afwijkend geboorte mechanisme (keizersnede of stuit bevalling) of familiale belasting. Echografie kon subjectieve klinische bevindingen als een positieve handgreep van Ortolani/Barlow objectief vastleggen. In dit onderzoek bestond een dergelijke klinische bevinding bij drieëntwintig heupen direct na de geboorte. Vijf van deze heupen waren echter stabiel bij het echografische onderzoek. Deze waren stabiel bij klinisch en echografisch onderzoek na een week, en zijn niet behandeld. Zij hebben zich gedurende hun eerste tien levens maanden normaal ontwikkeld, met een normaal klinisch, echografisch en röntgenologisch beeld. De heupen die ook volgens het neonatale echografische onderzoek geluxeerd of luxabel waren (achttien heupen) waren dit ook nog na één en na drie weken en zijn behandeld. Mogelijk zijn de heupen die echografisch stabiel waren bij het neonatale onderzoek voorbeelden van een fout-positieve uitslag van het klinische onderzoek. Echografie zou er toe bij kunnen dragen dat deze in wezen normale heupen niet worden behandeld.

Echografische instabiliteit (gedefinieerd in hoofdstuk V.4) in klinisch normale heupen kwam voor bij acht pasgeboren jongens en tweeëntwintig pasgeboren meisjes, in drieënveertig heupen. Alpha hoeken varieerden van 37-73 graden. Deze kinderen zijn opnieuw onderzocht op een gemiddelde leeftijd van 240 dagen, zonder te zijn behandeld. Ten tijde van het na-onderzoek waren al deze heupen echografisch stabiel, en had-

den zich klinisch, echografisch en röntgenologisch normaal ontwikkeld.

Bij het na-onderzoek is een vergelijking gemaakt tussen het echobeeld en de röntgenfoto. Aan dit deel van de studie zijn vijftientig andere zuigelingen toegevoegd, die waren verwezen wegens de verdenking op heupdysplasie. Drie heupen hadden normale alpha hoeken op het echobeeld, maar waren dysplastisch volgens de röntgenfoto. Het totale echobeeld van twee van deze drie heupen was wel afwijkend, met een afgeronde benige hoek en verbreed kaakbenig model. De acetabulum index lag tussen één en twee standaard deviaties boven de gemiddelde waarde volgens Tönis en Brunken (1968). Het röntgenonderzoek bevestigde een echografisch aangetoonde dysplasie in elf gevallen. Indien het röntgenonderzoek op deze leeftijd wordt beschouwd als de 'gouden standaard', dan is de sensitiviteit van echografie in dit onderzoek 78,6%, de specificiteit 99,4%, de voorspellende waarde van een positieve test uitslag 91,7% en van een negatieve test uitslag 98,3%.

Conclusies:

- Echografie dient niet te worden uitgevoerd als een algemene neonatale screening op congenitale heupdysplasie in klinisch normale pasgeborenen zonder bijkomende risico factoren. Hoewel een grote variatie in heup typen bestaat bij deze kinderen, met verschillende mate van steilheid van het acetabulum bij de geboorte, ontwikkelen zij zich haast altijd normaal.
- Echografische instabiliteit bij de geboorte in klinisch normale heupen is een onschuldige bevinding die niet leidt tot dysplasie. Het treedt frequenter op bij meisjes dan bij jongens.
- Echografie is waardevol om een positieve handgreep van Ortolani of Barlow te bevestigen.
- Pasgeborenen met risico factoren voor congenitale heupdysplasie zoals een stuit ligging, keizersnede of familiale belasting moeten regelmatig klinisch en echografisch worden gecontroleerd, daar zij gedurende hun eerste levens maanden een dysplasie kunnen ontwikkelen ondanks een normaal neonataal klinisch onderzoek.
- In vergelijking met röntgenonderzoek op de leeftijd van zes maanden heeft echografie een goede sensitiviteit en een uitstekende specificiteit, in het bijzonder wanneer het totale beeld (en niet slechts de alpha hoek) wordt beoordeeld.

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