VENOUS OUTFLOW OF THE BRAIN IN SPINA BIFIDA APERTA

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PROEFSCHRIFT

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CHAPTER I

INTRODUCTION

Children with untreated hydrocephalus show marked distension of the veins of the skull and scalp. This is even more pronounced in children with bifida aperta.

Such distension is usually relieved instantaneously when the hydrocephalus is treated by a ventriculo-cardiac drainage. This drew attention to the venous drainage of the cranial cavity and led to consideration of the possibility that impeded venous drainage might be a causal factor in the origin of hydrocephalus.

Initially, we examined brains of hydrocephalic children regardless of the cause of the hydrocephalus. After finding severe abnormalities of the cranial venous system in children with spina bifida aperta, however, we eventually confined our investigations to these cases. This limitation had the added advantage that we were dealing with a group of hydrocephalic children with the same aetiology and representing the single most common group available. Other forms of hydrocephalus have heterogenous causes that are even less clearly understood.

CHAPTER II

REVIEW OF THE LITERATURE

1) General remarks

The essential feature of hydrocephalus seen in children with spina bifida aperta, is the dilatation of the ventricles caused by cerebrospinal fluid (CSF) under increased pressure and a consequent cortical atrophy. It usually occurs at a time when the skull sutures are not closed, and therefore leads to excessive skull growth. It should be distinguished carefully from abnormal collections of fluid that are not under pressure, such as occur in porencephalic cysts within the brain substance or in enlarged cerebrospinal fluid pathways occupying space created by cerebral atrophy. In neonates and young children, progressive hydrocephalus need not be accompanied by brain atrophy. The cerebral hemisphere in newborn babies with spina bifida aperta may even be heavier than in normal babies (Emery 1964).

In some children arrest of progressive hydrocephalus occurs. The conditions for this arrest are still obscure, but apparently an equilibrium is established between the production and resorption of CSF. The labile equilibrium may, however, be disturbed, sometimes temporarily, by trauma or an infectious disease.

It is assumed that the CSF is mainly produced by the choroid plexus within the ventricular system and by the ependyma covering the ventricular wall. The production of CSF by the choroid plexus seems to be an active secretion. This secretion is probably not influenced by the intraventricular pressure. The fluid flows through the

ventricular system and out into the basal cisterns, from which it spreads into the subarachnoid spaces, around the brain and spinal cord.

The mechanism underlying the transport of the fluid to the blood is still not completely understood. Although in adults it probably occurs via the arachnoid villi and through the walls of the capillary bed of the pia-arachnoid, these villi are not yet demonstrable in the fetus, the neonate, or the very young child. In addition to being absorbed from the subarachnoid space, CSF seems to pass through the ependymal wall into the subependymal extracellular space, from which it returns to the capillary blood. The transmission of the fluid through the ependyma seems to be enhanced by small clefts present in the ependyma, especially under ventricular dilatation due to increased pressure. In cases of progressive hydrocephalus this capacity for resorption must be considerable, since it would otherwise be difficult to explain why the abnormal skull growth often slowly progresses over a period of weeks or months.

Disturbance of CSF circulation within the ventricular system is sometimes found in children with spina bifida aperta. Atresia or more often forking (Russell 1949) of the aquaeduct is a common finding (MacFarlane and Maloney 1957). In other children the exit of the fourth ventricle may be obstructed. Furthermore, the basal cisterns may be blocked. Another group of hydrocephalic patients seems to show a disturbance of the drainage from the subarachnoid space to the bloodstream.

In the normal embryo changes occur in the velum medullare posterius during formation of the extracerebral fluid pathways. The thin membrane is perforated, possibly as the result of the pressure of the fluid in the primitive ventricular system, which contains a functioning choroid plexus. According to the standing hypothesis the fluid then seeps into the perimedullary and extracerebral mesenchymal syncytium, clearing a way through this tissue in the course of the formation of the cisterns and other subarachnoid spaces. During subsequent development, the central canal of the spinal cord in the embryo becomes smaller and ceases to be important in the circulation of CSF. According to an old theory, this would not be the case in the presence of spina bifida aperta. In the ulcerating type, where a fistula is present, CSF is discharged. In the cystic form, resorption of CSF by the wall of the cyst can occur. The frequent occurrence of

hydromyelia in these children supports the theory that drainage of fluid by the central canal may modify the intracerebral pressures such that the normal extracerebral CSF pathways do not develop. The fact that skull growth increases rapidly after surgical treatment of the spina bifida aperta gives additional support to this hypothesis. Possibly, the absorption of CSF pulsations by the cystic spina bifida aperta, can mitigate the dilatation of the ventricles.

Gross abnormalities in the central nervous system are associated with spina bifida aperta. In particular, a complex abnormality of the medulla oblongata, cerebellum, and spinal cord was first reported by Cleland (1883), and was extensively described in 1891 and 1896 by Chiari.

Arnold's publication (1894) did not add further information to Chiari's accurate description. The Chiari II malformation, which was later named the Arnold-Chiari malformation by Arnold's assistents (Schwalbe and Gredig 1907), consists of displacement into the canal of the cervical spine of the cerebellar tonsils (occasionally with other parts of the cerebellum as well) and the obex of the fourth ventricle with the medulla oblongata. Sometimes, even parts of the pons are found within the cervical spinal canal. The malformation is accompanied by an elongation of the caudal part of the fourth ventricle. Because the cervical medulla is fixed by the denticulate ligaments, backward kinking occurs. The Cleland or Arnold-Chiari malformation may prevent passage of CSF to the basal cisterns and to the spinal canal, which leads to elevation of the intaventricular pressure.

In extreme pressure changes the Cleland or Arnold-Chiari malformation may function in the upper part of the cervical spinal canal as a ball-cock, thus producing intermittent obstruction of the foramen magnum. In fetuses with spina bifida aperta the persistence of the velum medullare posterius and the failure of the extracerebral CSF pathways to form may be due to the drainage of CSF through the central canal. This may not be the case in aquaeductal stenosis, which consist mainly of so-called forking. In this malformation the aquaeduct is represented by two or more distinct channels in normal nervous tissue. These channels are lined with ependymal epithelium. In addition, islands of ependymal cells are often seen in the neighbouring tissue. True atresia does not exist in these cases, since CSF can be drained through the separte small channels.

At ventriculography it may prove impossible to advance air through

the aquaeduct. A number of these children have forking of the aquaeduct at necropsy.

The cause of congenital hydrocephalus is usually sought in an obstruction at some point in the CSF pathways. Over-production of CSF in the presence of a normal rate of absorption has been considered in the rare cases of choroid plexus papilloma.

Diminution of the normal rate of CSF resorption with subsequent hydrocephalus due to chronic obstruction of the venous drainage of the brain has been assumed since the beginning of this century. Initial attemps to prove the assumption failed because of the abundant collateral venous circulation, but lately, evidence affirming the assumption has been collected.

2) Experiments on animals

Quite early, Dandy and Blackfan (1914) attempted to establish a relationship between intracranial venous pressure and hydrocephalus. They occluded the vein of Galen, or the straight sinus, or both, in ten dogs, only one of which developed an internal hydrocephalus. The other animals showed no signs of ventricular enlargement. The single instance in which hydrocephalus resulted was ascribed by the authors to the low placement of the clip at the origin of the great vein of Galen, thus preventing the principal tributaries from taking part in the formation of a collateral circulation. There was a distinct hydrocephalus with perforation of the septum pellucidum. The aquaeduct of Sylvius was also larger than normal. According to Dandy and Blackfan: ". . . . it was therefore evident that a low obstruction of the great vein of Galen may result in the production of an internal hydrocephalus, but that a high ligation had no such effect".

Guleke (1930) coagulated and cut the great vein of Galen at its origin, as Dandy and Blackfan suggested, in 9 dogs. Only one case of moderate ventricular dilatation was seen 2½ months after operation. Guleke did not think that ligation of the great vein of Galen was sufficiënt to give rise to hydrocephalus. He thought that additional circumstances must be present, but did not specify their nature.

Therefore there is reason to believe, that in both these series the dilatation of the cerebral ventricles was merely coincidental and not the result of the occlusion of the great vein of Galen. There is con-

siderable variation in canine ventricular size, and ventricular dilatation without clinical symptoms of intracranial hypertension can be found in an unusually high incidence in some breeds (Cammermeyer 1961).

Later investigators who performed similar experiments were unable to produce an experimental hydrocephalus in this way (Bedford 1934; Schlesinger 1939 and 1940; Hammock, Milhorat, Earle, and Di Chiro 1971).

Bedford (1934) reported on the results of obstruction of the great vein of Galen in 28 dogs. The animals remained in good health until they were killed three or four months later. In 13 cases the vein had been obstructed at its origin, and in 5 others comparatively close to its origin. None of these developed hydrocephalus.

Schlesinger (1934 and 1940) found hyperaemia in certain parts of the brain stem and of the hemisphere in all five Rhesus monkeys and one rabbit in which the small vein of Galen had been coagulated. In one monkey small haemorrhages occurred in the caudate nucleus on the side of the operation, but there was no hydrocephalus.

A study was done in a larger series of Rhesus monkeys and dogs by Hammock *et al.* (1971). The great vein of Galen was ligated in sixteen monkeys and six dogs, and the animals were followed for 6 months. Ten other dogs had originally been included in the experiments, but their pre-operative pneumoencephalograms showed large ventricles and they were discarded. None of the 22 animals in this study developed hydrocephalus. Postoperative angiograms showed dilatation of major draining veins and sinuses, and this persisted to some degree as long as the animals were followed.

Microscopically, there was pronounced dilatation of the smaller diencephalic and choroidal veins normally draining to the Galenic system. Ischaemic or atrophic changes could not be detected.

Further attempts to produce hydrocephalus secondary to venous blockage were made by Beck and Russel (1946), Bering and Salibi (1959) and Guthrie, Dunbar, and Karpell (1970).

Beck and Russell (1946) carried out experiments on the superior longitudinal sinus. Their attempts to produce sinus thrombosis just anterior to the torcular Herophili were surprisingly disappointing. Injection of thrombin or a chemical coagulant (an aqueous solution of 5 percent ethanolamine oleate with 2 per cent benzyl alcohol, which was frequently used in the treatment of

varicose veins) failed to cause thrombosis in four and nine rabbits respectively. Injection of the same chemical coagulant into three dogs failed go give any abnormality in the sinus either. Only microscopic damage to the sinus wall was seen after coagulation by heat in two dogs, and the vessel remained patent. Mechanical blockage by muscle in two dogs showed that the tissue was converted into firm greyish-white material narrowing the sinus considerably, but microscopical examination showed extensive recanalization after the dogs were killed, one 4 and the other 10 weeks later. Muscle steeped in a chemical coagulant produced a similar effect in four dogs.

Cottonwool steeped in chemical coagulant produced complete blockage only; no thrombosis ensued. In a litter of four puppies and one of four kittens the superior longitudinal sinus was succesfully blocked with cottonwool steeped in a chemical coagulant, but thrombosis of the sinus was not seen in any of these animals. The animals remained well and healthy until killed between 5 and 7 weeks later. The brains of the kittens showed no abnormality. In one puppy there was cerebral oedema and engorgement of the superficial veins.

Of the remaining three puppies two showed slight dilatation of both lateral ventricles and in the third the left ventricle was slightly more capacious than the right. On the basis of Cammermeyer's (1961) publication, the slight degree of dilatation observed in the puppies may be insignificant. In any case the blockage of the superior longitudinal sinus was not followed by any unequivocal clinical or pathological evidence of either increased intracranial pressure or hydrocephalus. Another noteworthy feature of these results is the resistance of the superior longitudinal sinus to thrombosis.

We now come to a second group of experiments on animals. Pressure studies have been done mainly in the dog, and it is difficult to judge whether the findings are also applicable to the human. The venous system in particular shows marked differences.

It must be kept in mind that the venous drainage of the cranial cavity is not the same in all species. In one form, shared by man, the Rhesus monkey, the pig, and the cat, the blood leaves the head through the jugular foramina via the internal jugular veins. But in the dog, horse, rabbit, sheep, and cow, the intracranial blood is drained via dorsal cerebral veins running through wide temporal foramina, and further drainage occurs via the external jugular veins. In the

latter group of animals the blood drained by the internal jugular veins comes from the pharynx, thyroid gland, and oesophagus, and there are few functioning shunts with cerebral vessels (Hegedus and Shackleford 1965).

Becht (1920) made a detailed study of the relationship between cerebrospinal fluid pressure, arterial pressure, and venous pressure in dogs, and came to the conclusion that an increase or decrease in the venous pressure, measured in the torcular Herophili, was rapidly and almost simultaneously followed by a corresponding increase or decrease of the CSF pressure, which was measured in the cisterna magna. He also found that an increase or decrease in arterial pressure, measured in the carotid artery, was followed by a rise or drop, respectively, in the CSF pressure, paralleled by similar changes in the venous pressure in the torcular. Lastly, he observed that a rise in the CSF pressure at a constant arterial pressure was not followed by a corresponding rise or drop in the venous pressure measured in the torcular. But when the CSF pressure approached the level of the diastolic blood pressure and the arterial pressure remained constant, the venous pressure dropped. Becht's findings all represent acute effects and no conclusion can be drawn concerning the relations of intracranial fluid pressures when long-lasting alterations occur.

As already mentioned, Becht concluded that elevation of the CSF pressure did not lead to elevated venous pressure in the torcular. This finding was confirmed by Weed and Flexner (1933), but is probably only applicable to the venous pressure measured in the dural sinus. Wright (1938) and Bedford (1942) found that the venous pressure measured in the posterior part of the superior longitudinal sinus dropped between 10 and 20 mm water after a slow or rapid increase of the fluid pressure, respectively. They explained this observation on the basis of impeded drainage via the great bridging veins to the sinus, i.e. in the region where the veins traverse the subarachnoid space. However, after this slight initial drop the venous pressure remained unaffected by further elevation of the CSF pressure.

In general, it holds that a normal superior longitudinal sinus and lateral sinus, with the triangular structure determined by the position in the junction of three dural layers, cannot be compressed. This explains why in this rigid structure the pressure is not influenced by the CSF pressure. If the same held for the other cerebral veins, it would hardly be possible for blood to leave the brain when the CSF pres-

sure is elevated.

Although the dural sinus pressure is only slightly, and then not correspondingly affected by the ventricular CSF pressure, a clear relation does exist between the CSF pressure and the pressure in the cerebral veins. Noell and Schneider (1948) isolated part of the longitudinal sinus such that it did not communicate with the sinus system. Pressure recordings in this portion of the longitudinal sinus reflected the pressure in the cerebral veins draining into the sinus. They demonstrated a parallel rise of the pressure in the cerebral veins and the CSF pressure in the great cisterns. This means that there must be a drop in pressure at the site of the entry of the cerebral veins into the sinus system. This decrease could be due to the anatomical configuration of the entry itself. It could also be explained by an empediment to slow the blood through the large draining veins in their course through the subarachnoid space toward the sinus by squeezing of the veins during pressure fluctuations.

Only Bering and Salibi (1959) succeeded in causing internal hydrocephalus in dogs by elevation of the intracranial venous pressure. After ligation of the internal and external jugular veins in the neck both the CSF pressure and the longitudinal sinus pressure were elevated, and hydrocephalus developed. The maximum of the ventricular dilatation was reached two to three weeks after the operation. Thirteen of the 21 dogs, which were examined, developed hydrocephalus, and eight did not. Of the latter one was killed the day after ligation and another was considered to have an incomplete ligation. The remaining six had complete ligation and developed elevated CSF and longitudinal sinus pressure, but did not develop hydrocephalus. The reason for this was not clear.

At autopsy no direct damage to the brain was found. The ventricular enlargement was thought to be caused by the possible failure of CSF absorption in the face of increased sinus pressure, and by the increased intraventricular pulse pressure from the choroid plexus. The reason that intracranial pressures dropped and ventricular dilatation ceased about three weeks after operation, was thought by the authors to lie in the development of a sufficiënt number of collaterals at this time. These collaterals were demonstrated subsequently on sinograms.

In experimental hydrocephalus obtained by injection of kaolin (aluminium stearate) into the cisterna magna of seventeen dogs,

Shulman, Yarnell, and Ransohoff (1964) found a greater increase of the longitudinal sinus pressure than of the CSF pressure. In twenty-four normal dogs these values were 90.3 ± 31.2 mm. H_2O and 147.1 ± 37.4 mm. H_2O respectively and in the 17 hydrocephalic dogs 204.9 ± 87.9 mm. H_2O and 207.4 ± 108.9 mm. H_2O, respectively. No further anatomic data are given, and no sinography was performed. The only possible conclusion which one can draw is that in kaolin-induced hydrocephalus longitudinal sinus pressure rises and the sinus pressure approaches the CSF pressure.

Lastly, Guthrie, Dunbar, and Karpell (1970) blocked the torcular Herophili and both lateral sinuses in 10 dogs by packing cotton pledgets into the bony canals through which the veins pass. Post-operatively, the superior longitudinal sinus pressure was measured at intervals of 1 to 29 weeks. The values for one dog were discarded because of faulty technique. There was a significant rise of the pressure. The mean pressure was 191 ± 113 mm. H_2O with a range of 72 to 471 mm. H_2O (pre-operatively 94 ± 33 mm. H_2O with a range of 55 to 177 mm. H_2O). In the 10 dogs the mean CSF pressure over the 29 weekperiod was 295 ± 189 mm. H_2O post-operatively, as compared to a control CSF pressure of 117 ± 54 mm. H_2O. The CSF pressure ranged from 79 to 742 mm. H_2O post-operatively.
The superior longitudinal sinus pressure and CSF pressure dropped, but were elevated for as long as 29 weeks. Superior longitudinal sinograms were performed post-operatively on 7 dogs. The development of collateral circulation around the torcular block was seen on the venous sinograms, explaining the gradual lowering of the superior longitudinal sinus pressure and CSF pressure. After torcular block the superior longitudinal sinus pulse wave was increased in amplitude, arterial in form, and synchronous with the pulse recorded in the lateral ventricle. This can be explained by the fact that the superior longitudinal sinus was isolated from the rest of the sinus system and the jugulars, and now expressed the intracerebral venous pressure wave. The resulting situation resembles the conditions prevailing in the experiments of Noell and Schneider (1948). The authors conclude that, although, the intracranial venous pressure was increased there seemed to be no change in the size of the ventricles. Their assumption is debatable, because they measured pressures in an isolated part of the sinus system and applied their findings to the whole cranial venous system.

Furthermore, only one measurement was performed in each animal and generalized for the whole group.

3) Investigations on human being

Initially, the possibility of a connection between elevated intracranial venous pressure and hydrocephalus was inferred from reports of cases with cerebral sinus thrombosis and ventricular enlargement. This led even Dandy and Blackfan to perform their experiments on the great vein of Galen and the straight sinus. From the review of these articles by Bedford (1939) we may quote the following: "The subject of Browning's excellent paper (1887) was a girl, aged 6, who died in coma after an illnes of three months. The symptoms were vague and indefinite. Autopsy revealed a dilatation of both lateral chambers, of the third and fourth ventricles and of the connecting iter; the straight sinus contained an organizing thrombus which, if not entirely blocking its lumen, very nearly did so. The thrombosis was the end-result of a perisinusitis of long standing. It will be observed that so far the case is beyond criticism. When, however, Browning came to examine the superior longitudinal sinus he found that although its lumen was everywhere free from antemortem clot, the walls of the sinus itself were involved in a perisinusitis similar to that affecting the straight sinus but which on naked-eye examination did not appear to be of as long standing as that of the latter. The lacunae laterales were especially affected and seemed for the greater part to have been reduced to fibrous cords. Unfortunately Browning made no histological investigations and thus we lack more precise details of the extent and relative duration of the lesions and also of the condition of the subarachnoid space generally. Thus the case is by no means free from complications and cannot therefore be admitted as an example of hydrocephalus following thrombosis of the straight sinus.

Newman's paper (1882) unfortunately lacks the wealth of detail which is to be found in Browning's. The patient was a man, aged 55, who died in coma after an illnes of about three months' duration. In vagueness of symptoms the case somewhat resembled the previous one. At autopsy the lateral ventricles were found to be considerably dilated and each contained an accumulation of about 4½ oz. of clear strawcoloured fluid which on examination was found to contain 0.09

per cent. of "albuminous matter"; microscopical examination of the sediment revealed a "few leucocytes and one or two crystals of cholesterine and margarine". The choroid plexuses were unusually vascular and there was a small whitish adherent clot at the point where the great vein of Galen and the inferior longitudinal sinus unite to form the straight sinus. None of the other vessels or sinuses contained clot. Unfortunately Newman failed to record the condition of the remaining ventricles and of the aquaeduct. It is impossible, therefore, to be certain that the case was one of communicating hydrocephalus."

Thrombosis of the dural sinuses in infants under three years of age is frequently seen when the child dies of a wasting disease. Thrombosis of the lateral sinuses resulting from inflammation in the middle ear and mastoid region, does not have as serious effects. Extension of the thrombus to other dural sinuses, or longitudinal sinus thrombosis, associated with meningitis or by other causes may have very serious effects. In a series of 80 such cases (Bailey and Hass 1937) 77 patients died with more or less extensive cerebral softening and necrosis with intracerebral and often subarachnoid haemorrhages. Three patients survived the acute illness to die later (Bailey and Hass 1937). There were a 6-year-old boy, a 10-month-old girl, and a boy 3½ months old at death, who had died 5 months, 9 months, and 2½ months after the onset of the illness respectively. The first child had acute pharyngitis and otitis media, and developed signs of intracranial hypertension with bilateral papilloedema. The second and third children had severe nutritional disturbances at the time of the onset of the intracranial disease. The second child had papilloedema as well, the third child did not, and his head circumference did not increase abnormally. The first and second children had subdural effusions. Only the third had gross cerebral atrophy. All had organized and canalized thrombi in the superior longitudinal sinus. The ventricles were moderately enlarged in the first two, the third showed gross ventricular dilatation, cerebral atrophy, and wide subarachnoid space over the convexity of the brain.

A case of superior longitudinal sinus thrombosis followed by hydrocephalus was reported by Ellis (1937) in an infant that developed multiple thrombosis following jaundice at the age of 5 weeks. The CSF was blood stained and under increase pressure. The anterior fontanelle was tense and there was slight separation of the

sutures. Gradual enlargement of the head was subsequently observed. Ventricular dilatation was demonstrated by air-ventriculography. Blockage of the superior longitudinal sinus was demonstrated radiographically following injection of positive contrast into the sinus at the site of the anterior fontanelle.

In her monograph on hydrocephalus, Russell (1949) discussed a possible relationship between dural sinus thrombosis and hydrocephalus. She suggested that sinus thrombosis may be followed by a limited degree of internal hydrocephalus after a certain length of time, perhaps a few weeks, and that the size of the ventricles may revert to normal, or almost normal, with canalization of the thrombus.

She reported the case of a 6-month-old infant who had had a discharge from the right ear six weeks before death. There was head enlargement (to 49 cm circumference), with bulging of the fontanelle. At necropsy great dilatation of the lateral ventricles, conspicuous haemorrhagic softening of the walls of both lateral ventricles, and similar softening in the basal ganglia was found. There were thrombi showing early organization in the superficial cerebral veins. Fresh antemortem thrombosis was found of superior longitudinal sinus, straight sinus, great vein of Galen.

Another case of a 15-year-old boy with presumable marantic sinus thrombosis was discussed in the same monograph. The boy had papilloedema, the left pupil was larger than the right, and there was bilateral abducens paresis. He had a hemiparesis on the left side. Further examination revealed an extensive tuberculous infection, Death was due to perforation of a tuberculous ulcer of the ileum. Organizing thrombus occupied the whole length of the superior longitudinal sinus and of the right lateral sinus in continuity. An area of haemorrhagic infarction was found in the right hemisphere. The ventricles were slightly and symmetrically dilated, including the third ventricle. The aquaeduct and fourth ventricle were not dilated and the foramina in the latter were patent.

Russell adds one other case of a female aged 20, who was admitted with a hemiparesis and who died of bronchopneumonia and focal encephalitis of mid-brain, pons and hypothalamus. There was a recanalized thrombosis of the lateral sinuses and the superior longitudinal sinus and a slight symmetrical dilatation of the lateral ventricles.

Thrombosis of the superior longitudinal sinus is presumably

related to haemoconcentration, decreased circulatory volume and flow rate, and local conditions of the vessels. It can be associated with malnutrition and prostration, and it can be secondary to infection. The usual symptoms are vomiting, diarrhoea, and dehydration followed by focal seizures, coma and death. The blood tinged and xanthochromic CSF with marked elevation of protein and a relatively small increase in cells may lead to the clinical impression of subarachnoid haemorrhage. When the patients die in the acute phase, not only thrombosis in the sinus system and superficial veins, but also brain swelling sometimes accompanied by uncal and tonsillar herniation is found. Subarachnoid haemorrhages are often noted over the convexities of the cerebral hemispheres as a consequence, of which perivascular and intracerebral haemorrhages and infarction in the cerebral cortex and white matter may be present. In addition, haemorrhage around subependymal veins, in the subependymal tissues, and within the lateral ventricles may be present. These extensive alterations, which are the result of sinus thrombosis may give rise to cerebral atrophy and scar formation, and CSF circulation disturbances may occur. It is difficult to assume that elevated cerebral venous pressure and disturbed CSF resorption through the pia-arachnoid villi are the sole reason for a possible ventricular dilatation when the child survives dural sinus thrombosis. In some cases cerebral atrophy and in other cases disturbance of CSF circulation through the subarachnoid space are likely to be major causes for ventricular dilatation.

Two children with gross hydrocephalus and formation of a bony ridge overlying the superior longitudinal sinus suggesting an aetiologic relation between superior longitudinal sinus pathology and hydrocephalus, were reported by Emery and Zachary (1956). The first child who was three weeks premature, became ill, running a temperature of 40°C, and was lethargic. He was treated by the family doctor and recovered from the acute illness. In the course of the next 2 years he developed a symmetrical gross hydrocephalus and at the time of the ventricular studies the ventricular pressure and the lumbar pressure were more than 40 cm H_2O. The hydrocephalus was shown to be communicating, but it was not mentioned whether air passed to the subarachnoid space covering the convexity of the brain. He was treated with a spinal theca-ureter shunt, and died three weeks later of electrolyte disturbance due to unrestricted drainage of CSF

into the urinary tract. At necropsy, the superior longitudinal sinus appeared to be converted into a firm, fibrous mass containing a few small blood vessels. The skull showed a bony ridge immediately over the longitudinal sinus over the whole length of the vault. Within the fibrous mass into which the sinus had been converted, no blood pigment was found, and inflammatory signs were also absent. The second child had meningitis due to *B.coli* at 1 week of age. He was treated with antibiotics and appeared to recover completely. His head increased rapidly in size and he was admitted at 3 months of age. He was found to have a communicating hydrocephalus in which the ventricles were enormous. A spinal theca-ureter shunt was made. At three months of age he was well. On examination, a bony ridge dividing the anterior fontanelle into two lateral fontanelles was felt. In the first case the illness responsible for the sinus occlusion (possibly meningitis) must have taken place at 1 week of age, thus explaining why no signs of thrombosis were found at the time of death. The second case was comparable to the first in that a bony ridge was present over the sinus, but lacks the wealth of detail offered by the first case.

Kinal (1962) reported four children with hydrocephalus showing anomalies of the lateral sinuses. The sinograms of the first case disclosed stenoses of the left lateral sinus. The second child had stenotic sigmoid sinuses and left lateral sinus. The third child had an obliquely situated right lateral sinus and right stenotic sigmoid sinus and an obliquely situated stenotic left lateral sinus with atresia of sigmoid sinus and jugular bulb and vein. The fourth case showed a moderately narrowed right lateral sinus and stenotic sigmoid sinus. None of the reports mentions the cause of the hydrocephalus or whether a spina bifida aperta was present.

Shulman and Ransohoff (1965) investigated fifteen children with hydrocephalus; their paper does not mention whether these cases concerned children with spina bifida aperta or the cause of the hydrocephalus. The venous pressure in the superior longitudinal sinus and the CSF pressure were both elevated, the ratio between these pressures being 1.08 to 1. When fluid was allowed to drain from the ventricular system, the venous pressure dropped simultaneously but not below the level of the CSF pressure. Sinograms were performed in three instances. In two children no distinct anomalies were seen. In the third infant the lateral sinus on each side seemed to taper to

end proximal to the jugular foramen, and blood from the sinus was seen to be exiting via enlarged parietal and mastoid emissary channels.
The photographs pertaining to this case suggest that it concerned a child with spina bifida aperta. The ventricular CSF pressure was not measured during sinography.

In an investigation of eleven hydrocephalic children with spina bifida aperta and nineteen children with hydrocephalus from other causes, Norrell, Wilson, Howieson, Megison and Bertan (1969) demonstrated a parallel rise of the venous pressure in the superior longitudinal sinus and of the CSF pressure after infusion of saline into the lateral ventricle in the children with spina bifida aperta. This parallel rise did not occur in the other children. The sinography performed during the infusion provided the explanation of this phenomenon. During appreciably elevated pressure in the ventricular system, partial or complete obliteration of the lateral sinuses occurred, but this was never observed when the sinogram was performed while the CSF pressure was normal. Associated with obstruction of the blood flow through the lateral sinusses, the venous drainage took place via collateral routes. It was never found, however, that the venous pressure in the sinus system was higher than the CSF pressure in the ventricles.

In a number of cases too, elevated venous pressure may be due to arterio-venous aneurysms, which are consequently accompanied by ventricular dilatation. A detailed publication on arterio-venous aneurysms of the great cerebral vein of Galen with a comprehensive review to the literature on this subject by Gold, Ransohoff, and Carter (1964) indicates that the occurrence of hydrocephalus in patients with these aneurysms is common. Although compression of the mesencephalon with stenosis of the aquaeduct of Sylvius may well lead to hydrocephalus as the authors suggest, elevated venous pressure in the cerebral venous system may be the cause of or contribute to the ventricular dilatation.

Arterio-venous angiomas may give rise to hydrocephalus as well (de Lange and de Vlieger 1970). In this case an extracranial arterio-venous malformation with connections with the lateral sinus was reported. Elevated sinus pressure with high pulsations was recorded. After ligation of the external carotid artery, the sinus pressure fell and the height of the venous pulsations diminished from 50 to below

17 mm. H_2O. Air studies done pre-operatively and 8½ months later demonstrated a decrease of the ventricular size, most pronouced in the frontal horns.

A case of superior vena cava obstruction was discussed by Hooper (1961). The infant developed obstruction of the superior vena cava progressively in the course of 16 months. No clear cause was found, although hyperplasia of the thymus was found at operation. Angiography revealed no obstruction of the intracranial sinuses. A subsequent venous angiocardiogram revealed complete obstruction of the superior vena cava and of the azygos vein at its opening into the superior vena cava. The dye reached the heart via collateral routes. At the start of the illness his head was large, and the size increased. At the age of 9 months a pneumoencephalogram showed the lateral and third ventricles grossly dilated and the subarachnoid space filled. No clear abnormalities apart from the communicating hydrocephalus were found. The child was treated with a shunt and remained well.

Several otorhinolaryngologists have published CSF pressure studies based on unilateral or bilateral ligation of the jugular vein during block dissection in the neck (Sugarbaker and Wiley 1951, Schweitzer and Leak 1952, Royster 1953). After these operations the patients frequently complained of headache and in all the cases the lumbar puncture results showed elevated CSF pressure. The CSF pressure varied considerably, but was often higher than 60 cm. H_2O shortly after the operation. After a few weeks, however, the lumbar CSF pressure was found to be normal or slightly raised (25 cm. H_2O), probably because the drainage of blood from the head was facilitated by improvement of the collateral drainage. The patients who had pressures around 60 cm. H_2O complained of nausea and giddiness. It was not mentioned whether papilloedema had been present. Sugarbaker and Wiley (1951) also reported a rise of the lumbar CSF pressure a few weeks after bilateral resection of the internal jugular veins, upon compression of the cervical muscles. After the neck operation, collateral venous drainage was established via the vertebral column. The rise of the lumbar CSF pressure seems to have arisen because of compression of the external vertebral plexus, giving an effect in these patients analogous to that obtained in Queckenstedt's test.

4) **From the reports in the literature, the following assumptions relevant to the present study can be made.**
   1) Acutely, there is a parallel rise of the CSF pressure with the dural sinus pressure, but not of the dural sinus pressure when the CSF pressure is elevated.
   2) In experimental and clinical hydrocephalus there is an elevated dural sinus pressure.
   3) Elevation of the CSF pressure in the ventricular system leads to elevation of the dural sinus pressure in children with spina bifida aperta.
   4) Elevated intracranial venous pressure can cause hydrocephalus.

   The following postulates will be evaluated on the basis of the results and were either refuted or confirmed by the present study.
   1) Congenital anomalies of the intracranial venous system originate primarily as components of a disturbance in embryonic development, and can lead to elevated venous pressure, which in turn leads to hydrocephalus.
   2) The hydrocephalus already present in the intra-uterine phase leads to changes in the venous drainage of the brain, thus giving rise to what for man is an abnormal drainage via the external jugular system.
   3) Elevated CSF pressure in the ventricular system results, among other things, in elevated intracranial venous pressure, which helps to maintain the elevated CSF pressure and aggravates the brain damage due to hydrocephalus, thus constituting a vicious circle.

CHAPTER III

SOME FACTS OF VENOUS DRAINAGE OF THE NORMAL BRAIN AND IN SPINA BIFIDA APERTA (FROM THE LITERATURE)

1) The drainage of the cranial cavity

The blood leaves the cranial cavity via foramina which are completely or partially occupied by a vein. In some cases, e.g. the foramen magnum, the veins take only a limited amount of the space.

a. *The jugular foramina*

The jugular foramina lie on either side of the clivus where the posterior side of the petrosa borders on the occipital bone. They are composed of an anterior part, the pars nervosa, through which the glossopharyngeal, vagus, and accessory nerves run, and a posterior part, the pars venosa, traversed by the internal jugular vein, the most important vessel in the drainage of the cerebrum. The first accurate measurements of the jugular foramen are probably those made by Kasloff (1844, cited by Rüdinger 1876) who assumed a relationship between narrowing of the foramen and certain mental diseases. Since then, many papers on measurements of the jugular foramen have appeared, particularly in the German literature. Otorhinolaryngologists have also published on this object.

Hayek (1929) investigated the shape and variability in about 100 skulls. He found not only a wide range in the size and asymmetry between the right and left foramina but also that in many cases the pars nervosa and the pars venosa formed separate foramina.

Recently, Naga (1966) made a study of the jugular foramen and the bulb of the jugular vein. According to the literature, on the average the venous portion of the right foramen is larger than the left in 70 per cent of the cases, the left larger than the right in 20 per cent, and in 5 per cent the foramina are symmetrical.

b. *The emissary foramina*

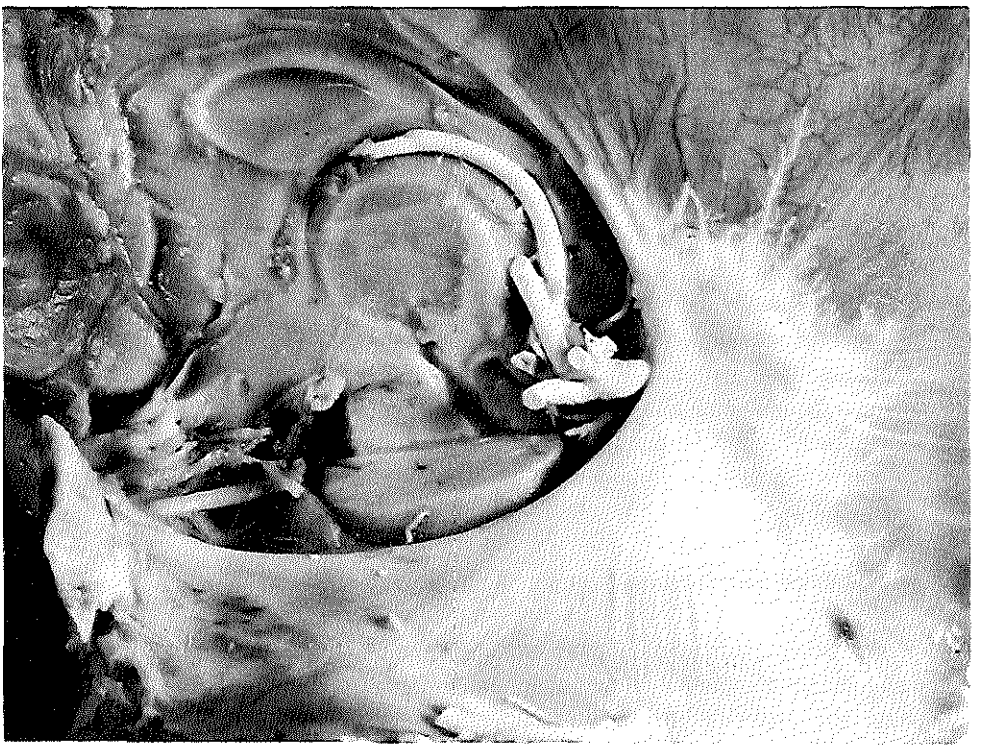
Although the drainage of blood via the jugular foramina is the most important, there are other routes whose individual contribution is difficult to estimate. When there is obstruction of the drainage via the jugular route in the widest sense, however, the combined share of all these veins must amount to an appreciable alternative system. One group of these accessories is formed by the emissary veins, which pass through the emissary foramina.

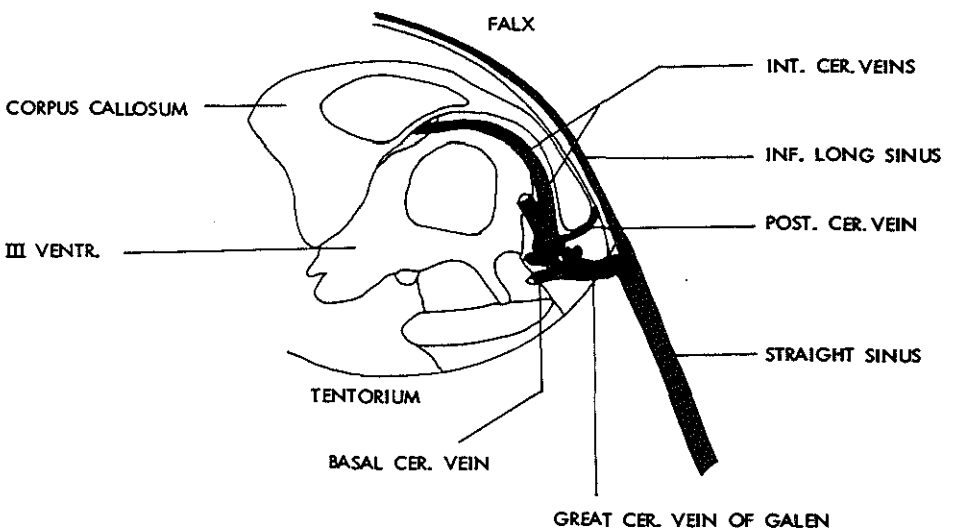
2) Deep veins and sinuses in the mid-line

The blood from the deep cerebral regions is drained mainly via the straight sinus, which traverses the tentorium at the site of attachment of the falx. Here it has a triangular structure. To reach the origin of the straight sinus, the blood passes through the inferior longitudinal sinus, the great cerebral vein of Galen, and the basal cerebral veins. The inferior longitudinal sinus runs within the free lower margin of the falx, and in the normal specimen seems to contribute little to the drainage of the brain.

The wide-calibred great vein of Galen is short in the normal specimen (Fig. 1a and b). This large vein is formed mainly by the internal cerebral veins, arising above the pineal gland and curving around the splenium of the corpus callosum toward the origin of the straight sinus. The internal cerebral veins run over the third ventricle in the arachnoid space on either side of the median line.

The vein arises from the septal vein running on either side of the septum pellucidum and arising in the interventricular foramen of Monro. The thalamostriate vein empties into the internal cerebral vein in the foramen. The vein runs over the floor of the lateral ventricle between the caudate nucleus and the thalamus, from latero-occipital to medio-frontal, and receives many rami from the anterior and middle parts of the wall of the lateral ventricle. The thalamostriate vein also





Figs. 1[a] and [b] In a normal head the straight sinus and deep cerebral venous system are injected. The left cerebral hemisphere has been removed. Compare the normal relationship of the vessels with the abnormal course taken in spina bifida aperta (Figs. 8 and 9).

receives veins draining the choroid plexus and the corpus callosum. A separate vein from the posterior horn also often empties into the thalamastriate vein. The posterior cerebral vein, which in the normal specimen has a rather small calibre, runs along the posterior side of the corpus callosum and curves around the splenium towards the great cerebral vein of Galen or to the origin of the straight sinus, into which it empties.

The basal cerebral vein arises near the perforate substance and above the anterior medial border of the temporal lobe, 1 to 1.5 cm. lateral to the optic chiasma. It then runs posteriorly and laterally around the uncus and approaches the lateral border of the cerebral peduncle. It continues in the cisterna ambiens, running posteriorly and upward medially, and empties into the vein of Galen (Johanson 1954).

3) Superior longitudinal sinus and ascending cerebral veins

During embryonic development, the primitive superior longitudinal sinus receives a larger amount of tributaries which are derived from the anterior dural plexus. During the elaboration of the sinus these veins are reduced and those veins that remain are rendered more conspicuous as they become larger and longer in traversing the arachnoidal layer. Eventually, there are only four to six major stems by which the ascending cerebral veins of each side enter the superior longitudinal sinus (Stopford 1930). The stems of these veins, for some distance before their sinus entrance, are either attached to the dura or lie in it, or are free in the subdural space, as is particularly noticeable in the infant (Padget 1956).

In the literature (Stopford 1930) importance is assigned to the direction from which the ascending cerebral veins enter the superior longitudinal sinus. This is reported to be frontal in the majority of the cases, which means that the veins empty against the direction of flow in the sinus and that might have haemodynamic implications. This frontal direction appears in late fetal life (Padget 1957). It can be the mechanical result of the backward expansion of the cerebral hemispheres.

During fetal development there is caudad migration of the primitive transverse sinus in the meshes of the tentorial dural plexus. At the midline, the cerebral hemispheres become approximated and the (primitive) marginal sinuses, which follow the dorsocaudal borders of

the hemispheres, join in a midline anastomosis. The superior longitudinal sinus emerges by a combination of two processes: the unequal or alternate enlargement of the channel of one side with a dwindling of the opposite vessel, and a coalescence of the bilateral channels.

4) Position of the torcular with respect to the foramen magnum

A remarkable feature of the brain of children with spina bifida aperta is the steep slope of the tentorium and the close apposition of the torcular to the margin of the foramen magnum (Cameron 1957, Daniel and Strich 1958). It was therefore of interest to determine whether the antero-posterior diameter of the foramen magnum was the same in these patients as in normals and, if so, whether the distance from the torcular to the anterior margin of the foramen magnum diverged appreciably in children with spina bifida aperta as compared with normal children. Kruijff and Jeffs (1966), who measured the foramen magnum on radiograms of 80 children with spina bifida aperta and compared the results with the findings in normal children in comparable age-groups, came to the conclusion that in children with spina bifida aperta the foramen was larger than that of normals in 71 per cent of the cases. However, these measurements did not determine the true dimensions, because the cartilaginous and fibrous parts of the foramen cannot be measured on radiograms and the ossification of the skull is not the same in spina bifida aperta patients as in normal children. Therefore, we performed our measurements in unmacerated material.

Drainage from (A) and via (B) the posterior fossa

A. On the basis of direction of drainage, the veins of the posterior fossa can be divided into three systems:

   a) A superior group

   or those vessels draining superiorly or deeply into the great vein of Galen. Included in this group are the posterior mesencephalic vein, which originates from the lateral or anterior aspect of the mesencephalon, the precentral cerebellar vein running in the precentral cerebellar fissure, and the superior vermian vein draining the antero-superior portion of the superior vermis and adjacent cerebellar hemispheres (Huang and Wolf 1965).

b) A posterior group
or those vessels draining posteriorly or laterally into the torcular and neighbouring straight and lateral sinuses. The inferior vermian vein originates on the cerebellar tonsils and runs in the median groove between the cerebellar hemispheres to the straight sinus. It receives tributaries from the cerebellar tonsils, pyramids, and tuber. Near its junction with the straight sinus it receives a tributary from the declive. Multiple hemispheric tributaries join the superior vermian vein in its course (Huang, Wolf and Okudera 1969). The upper aspect of the cerebellar hemispheres drain into sinuses running through the tentorium to the lateral sinuses.
c) An anterior group
or those vessels draining anteriorly into the petrosal sinuses. The petrous or petrosal vein drains the pons, medulla oblongata, and also the dentate nucleus. Via the brachial vein lying in the depth of the precentral cerebellar fissure, the petrosal vein is anastomosed to the precentral cerebellar vein. The petrosal vein empties into the superficial petrosal sinus.

B. In addition to a venous system draining the cerebellar structures in the posterior fossa, there is also a large drainage system in the dural layers, serving the intracranial cavity.
The dura of the posterior fossa usually – and always in young children – has an occipital sinus lying in the median line. It runs in the dura covering the posterior fossa where the cerebellar falx joins this structure. This sinus connects the torcular with the marginal sinuses lying around the foramen magnum communicating with the last part of the sigmoid sinus and the internal vertebral plexus.
On the clivus there is also the basal plexus providing communication with the plexus around the sella and the inferior petrosal sinus and having adequate contact with the vertebral plexus.

CHAPTER IV

MATERIAL AND METHODS

The study was performed in the Department of Pathology, Children's Hospital, and the Congenital Anomalies Research Unit, both in the University of Sheffield and in the University Hospital at Rotterdam.

The specimens derived from children with spina bifida aperta and children whose death was due to diseases not involving the head. No distinction was made in this study between meningocele, cystous meningocele, and the ulcerous forms. Pure meningocele did not occur in the series, probably because these children survived. The spina bifida was not localized in the cervical region in any of the cases.

The same method of preparing the specimens was used throughout. Twenty-four hours before autopsy, the ventricular cerebrospinal fluid was replaced by a 4 per cent formalin solution. After the general autopsy the brain and spinal cord were removed in their bony compartments and all tissues fixed in formalin before further dissection. The minimal duration of fixation was fourteen days, and the specimens were stored until required. This method has been routinely applied for many years in Sheffield, where it has provided a reserve of several hundred specimens from children with spina bifida aperta and/or hydrocephalus for research purposes. The method has been used in Rotterdam since 1968 and a series of more than twenty preparations is now available for study.

In a number of the specimens the intracranial veins were injected

with a 15 per cent aqueous suspension of colloidal barium sulphate (micropaque) and 5 per cent gelatine prior to the autopsy, with variable success.

In most of the cases injection was done via the right internal jugular vein in the neck after ligation of the common facial vein and of both internal jugular veins cardiac to the injection site. In a few cases the straight sinus was catheterized, via an opening (1 x 1 cm) made in the skull at the level of the torcular, for the introduction of the warm barium solution. This material was studied radiographically. In a few preparations the same contrast medium had been injected into the arteries for an earlier, unrelated study.

Technical details of methods required for the parts of the study are given below.

1. Jugular foramen

The venous portion of the jugular foramen was measured in 100 skulls of children with spina bifida aperta and 35 skull of children with affections not involving the nervous system. After dissection of the foramen, the smallest and largest diameters were measured with blunt-ended calipers and read with a vernier gauge to a tenth of a millimetre.

2. Deep veins and sinuses in the mid-line

Fifty-two preparations were used for this apart of the study. The ages varied from one day to three years and eleven months. After fixation, the skull was opened on the left side along the superior longitudinal sinus and the left transverse sinus. Dissection was performed in a sagittal plane along the falx and through the third ventricle, after which the cerebral peduncle was severed and the left hemisphere removed. The falx cerebri, the tentorium, and the blood vessels in the mid-line were left *in situ*.

3. Superior longitudinal sinus and ascending cerebral veins

In 28 specimens, 2 cm. of cranial bone was removed on either side of the sagittal suture, leaving the dura intact. The superior longitudinal sinus was cut open and then the dura 2 cm. lateral to the sinus. Cuts made at right angles to the sinus created dural flaps permitting observation of the ascending cerebral veins in their normal anatomical

relationships.

The junctions of the veins with the sinus were localized and the distance to the torcular measured in centimetres with a measuring tape, the origin of the torcular being taken as the point of entry of the straight sinus. The angle at which the veins entered the superior longitudinal sinus was also measured. The course of the ascending cerebral veins could not be described in relation to the sulcus centralis because the course of the sulcus could not be determined exactly. In cases where a structure resembling a sulcus centralis was discernible on the convexity, no continuation on the median cortex could be observed.

The ages at death in this series varied from one day to two years and eleven months.

4. The other descending supratentorial veins and sinuses

This part of the study was done in 30 specimens.

5a. Position of the torcular with respect to the foramen magnum

The antero-posterior and transverse diameters of the foramen magnum were measured with calipers and read with a vernier gauge in 57 skulls of children with spina bifida aperta and 14 skulls of normal children.

In 27 specimens from children with spina bifida aperta and in 13 skulls of normal children the distance of the torcular to the ventral edge of the foramen magnum was measured. The point of entry of the straight sinus into the torcular was taken as measuring point.

5b. Veins and sinuses in the tentorium and over the middle fossa

Fifty specimens were used for this part of the study. The skull was removed supratentorially. During the detachment of the brain, a careful search was made for venous connections with the basal venous plexuses.

5c. Drainage from and via the posterior fossa

This part of the study was done in 20 specimens.

CHAPTER V

RESULTS

1. Drainage of the cranial cavity

a. The jugular foramina

The jugular foramina were present in all the cases. The size of the foramina varied greatly in both the group with spina bifida aperta and the normal group; ranging from 1.8 mm to 8.0 mm. in children with spina bifida aperta and from 2.0 mm. to 7.0 mm. in normals. The right foramen was larger than the left in 68 of the children with spina bifida aperta, the left larger than the right in 20, and in 12 they were symmetrical. The size of the foramina in 104 children who died under the age of 6 month, both children with spina bifida aperta and normals, are shown in Table 1. In this diagram the vertical axis represents the sum of the various diameters. On casual inspection there would appear to be a slight difference between the readings of children with spina bifida aperta and those without, but statistical analysis showed no valid difference (Blaauw and Emery, 1969).

b. The emissary foramina

The *parietal foramina*, which lie on either side of the mid-line through the vertex, were present in all the cases and showed a diameter of 1.5 mm. to 3 mm. in the injected specimens. The *occipital foramen* seemed to have limited importance for drainage, and was missing in 60 per cent of the specimens. It was certainly not one of the major emissaries. A common form is shown in Fig.2: with the internal view on the left and the external view on the right, of the

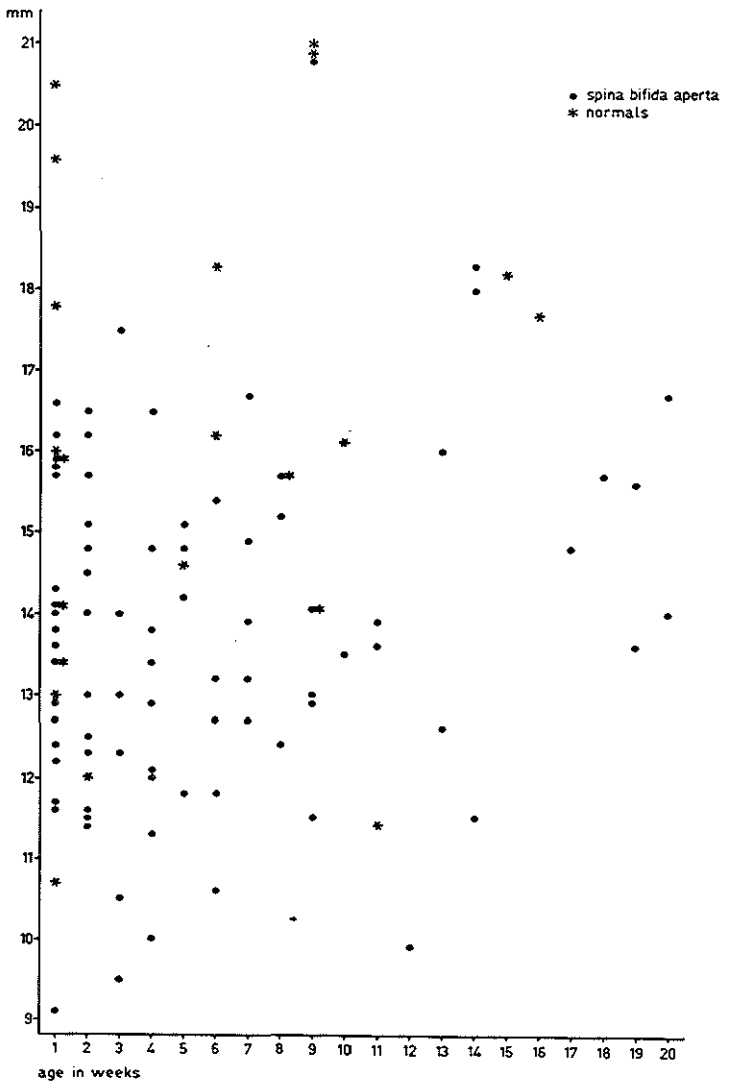


Table 1 Size of the jugular foramina in 104 children who died under the age of 6 months, both with and without spina bifida aperta. The vertical axis represents the sum of the various diameters.

occipital bone at the site of the protuberance and the location of the torcular. One or in some cases several vessels of the torcular communicated with diploic vessels situated in a thickening in the mid-line of the occipital bone. On the outer side, very small outlets of these vessels can be seen. In many cases, too, a vessel left the posterior third of the superior longitudinal sinus to form a shunt with these diploic vessels.

In the normal situation too, a *sphenoid emissary* runs through the foramen ovale. In this series of children with spina bifida aperta the veins passing together with the trigeminal nerve through the foramen ovale were often conspicuous. (Fig.3). In some cases the dural venous plexus of the middle fossa seemed to form only communication with the transverse sinus, but others were true tentorial sinuses, although with shunts to the transverse sinus. The sphenoid emissary then formed extracranial anastomoses with the pterygoid plexus.

c. The ophtalmic vein

The cavernous sinus showed limited dimensions in these specimens. Because the majority derived from very young children, it could not be determined with certainty whether this sinus increases in size and importance with advancing age. In any case the ophthalmic vein, judged at the point where it leaves the skull via the superior orbital fissure, seemed of minor importance as an anastomosis with the external jugular system. These findings were in good agreement with observations made in skulls of young children who had suffered from extracranial affections.

d. Venous drainage to the internal and external vertebral plexus

One of the striking findings in this study was the accumulation of contrast medium in the region of the cervical spine in the injected specimens. Although the jugular veins had been ligated proximally to the site of injection and the contrast medium was injected under very little pressure, the medium flowed out of the cranial cavity with ease and arrived via the vertebral plexus in the superior vena cava, often filling the heart, while little was found in the external jugular vein. Although this might be an indication of a very effective drainage pathway from the cranial cavity via the spinal column, it remains a question whether this had been the case *in vivo*.

From the posterior fossa the marginal plexus made a transition

into the internal vertebral plexus. In addition, emissary veins communicated with the external vertebral plexus via the hypoglossal foramen and the condyloid emissary veins. Large veins accompanied the nerve roots in the cervical spinal column, particularly C7 but also C6 and C8, anastomosing with the external vertebral plexus. In all the specimens only the usual emissaries were present.

The skull in spina bifida aperta shows the normal number and configurations of the jugular foramen and emissaries. The drainage via the vertebral column seems to be enhanced.

2. Deep veins and sinuses in the mid-line

Forty-eight of the 52 cases showed moderate to marked enlargement of the ventricles, most pronounced in the lateral ones.

a. Internal cerebral vein

The internal cerebral vein arose in the interventricular foramen and passed over the roof of the third ventricle to the pineal body, where it joined the corresponding vein from the other side and continued as the great cerebral vein of Galen. In eight cases there was an arachnoid cyst on the posterior and upper side of the third ventricle; these cysts probably represented a cavum velum interpositum. In all these cases the internal cerebral vein passed around the cyst laterally.

The septal vein is usually the first tributary of the internal cerebral vein. In the injected preparations it was insignificant and in the uninjected material sometimes even unrecognizable, possibly due to the hypoplasia of the septum pellucidum, which was sometimes virtually absent. The thalamostriate vein, on the contrary, was very pronounced, and its course long; it arose in the roof of the temporal horn and passed over the basal ganglia to the interventricular foramen, acquiring numerous tributary branches running subependymally (Figs.4, 5 and 6).

The posterior cerebral vein originated at the level of the middle of the often hypoplastic corpus callosum, around which it ran occipitally, and emptied into the internal cerebral vein or the origin of the vein of Galen. The posterior cerebral vein often showed shunts to the inferior and superior longitudinal sinuses via collaterals.

b. Basal vein

The basal cerebral vein originated at the base of the brain from perforating veins of the corpus striatum, and communicated with the cavernous sinus, the anterior cerebral vein, and the superficial petrosal sinus. It occurred in all the specimens and seemed to have a normal course with respect to the brain structures.

c. Great cerebral vein of Galen

The great vein of Galen arose, in spina bifida aperta as in the normal situation from the junction of the two internal cerebral veins, which was consistently located behind the pineal gland. The vein was, however, always several times longer in cases of spina bifida aperta than in the normal specimens, as mentioned by Daniel and Strich (1958). In one case it even reached a length of 12 cm. (Fig.7).

In the present series the vein of Galen showed two courses. In one, the vein ran from the pineal body over the cerebellum to the tentorium at an obtuse angle to the straight sinus (Fig.8[a] and [b]). In the other (Fig.9[a] and [b]), which differed widely from the first, it ran from the pineal body upward in the direction of the inion and then turned sharply downward in the caudal direction. The angle lay either in the tentorial-notch, so that the vein substituted for the straight sinus, or in the arachnoid space because the vein reached the tentorium and the inferior longitudinal sinus further down. The first of these patterns was seen in 22 cases, the second in 30 cases.

In a number of cases the great vein of Galen was partially or entirely duplicated (Fig.9[a]).

d. Straight sinus

The attachment of the falx to the tentorium was narrow. The tentorium was situated close to the foramen magnum, lying at a sharp angle to the base of the skull. Because of the short root of the falx and the steep slope of the tentorium, the straight sinus was also short and steep and sometimes seemed as it were to touch the occipital bone.

e. Falx cerebri and superior and inferior longitudinal sinuses

The falx was highly abnormal in all cases, being narrow and shallow. In 39 of the specimens the middle third was either absent or poorly developed and fenestrated. In 12 cases belonging to the

second group, the inferior longitudinal sinus ran in a band of dura (Fig.10). In one case both the anterior and middle thirds were absent, and in another the middle and posterior thirds were poorly developed.

When the falx was absent or narrow and fenestrated, the convolutions of the medial surfaces of the hemispheres were interdigitated and the pia-arachnoidal covering was fused. In about a fourth of the cases the course of the inferior longitudinal sinus was roughly normal, but in the other case it was often absent in the middle third of the falx. In these latter cases too, however, the rudimentary frontal portion was distinct.

In 40 specimens one sinus and sometimes several ran transversely through the falx from the free margin to the superior longitudinal sinus, always in the middle third of the falx at the level of the anterior fontanelle, and emptied into the superior longitudinal sinus (Figs.11 and 12). These collateral sinuses occurred independent of whether the inferior longitudinal sinus was normal or showed only a frontal rudiment. The tributaries of these sinuses came from the medial cerebral surface and showed anastomoses with the anterior and posterior cerebral veins. These dural collaterals are of particular interest because it is at this site that the superior longitudinal sinus communicates with the scalp veins and thus with the external jugular system via the parietal emissary veins.

In one case the inferior longitudinal sinus emptied into the superior longitudinal sinus above the torcular and independent of the straight sinus. In two children with spina bifida aperta but no gross hydrocephalus, the configuration was even more divergent. In the first specimen the tentorium was completely absent. The "torcular" was situated on the margin of the foramen magnum, and the transverse sinus lay in small dural folds around the foramen. The junction of the straight sinus and the inferior longitudinal sinus lay far above the "torcular" (Fig.13). In the second the torcular lay at the level of the inion and the transverse sinuses ran in long narrow leaves of the tentorium, enclosing the occipital lobes which consequently lay partially infratentorially (Figs.14[a] and [b], 15). In this last case the posterior cerebral vein emptied into the inferior longitudinal sinus, but just occipital to the junction there was a wide shunt between the internal cerebral veins and the inferior longitudinal sinus (Blaauw 1970).

The noteworthy deviations from the normal anatomy treated in this section is formed by the transverse collaterals in the falx at the side of the anterior fontanelle. There are also clear abnormalities of the falx and the inferior longitudinal sinus. The great vein of Galen/ straight sinus showed two courses. The great vein of Galen was several times longer. The straight sinus was short and took a more vertical direction.

3. Superior longitudinal sinus and ascending cerebral veins

In all the specimens used for this and other parts of the study, a superior longitudinal sinus was present. In one case the sinus was obliterated at the site of the anterior fontanelle; this seemed to be an artefact, possibly caused by punctures.

It was clear that the anterior third of the superior longitudinal sinus did not lie precisely in the junction of the parietal dura and the falx. The sinus ran frontally as far as the bregma, 1 to 3 cm. from the convexity in the falx.

Partitions of varying length were sometimes seen in the sinus, dividing it into two parts. In all cases these partitions ran in the same direction as the falx. Specimens were also seen in which the superior longitudinal sinus was duplicated, the second sinus lying parallel to the first in the falx, from which it arose and into which it emptied (Fig.16). In one case veins from the medial cortex emptied into this parallel sinus. In another case wide ascending cerebral veins curved over the medial cortex and continued in the falx, running parallel to the longitudinal sinus to the torcular, and emptied about 2 cm further on in the floor of the longitudinal sinus.

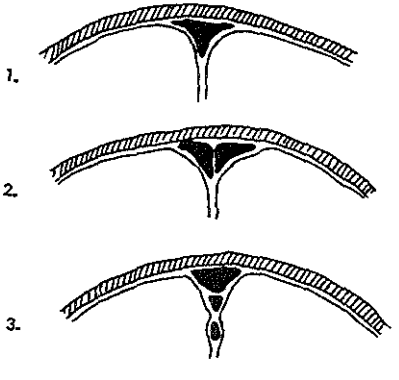


Fig.16 Drawing representing a transverse section through the longitudinal sinus (1), a sinus with partitions (2), and a sinus which is duplicated (3).

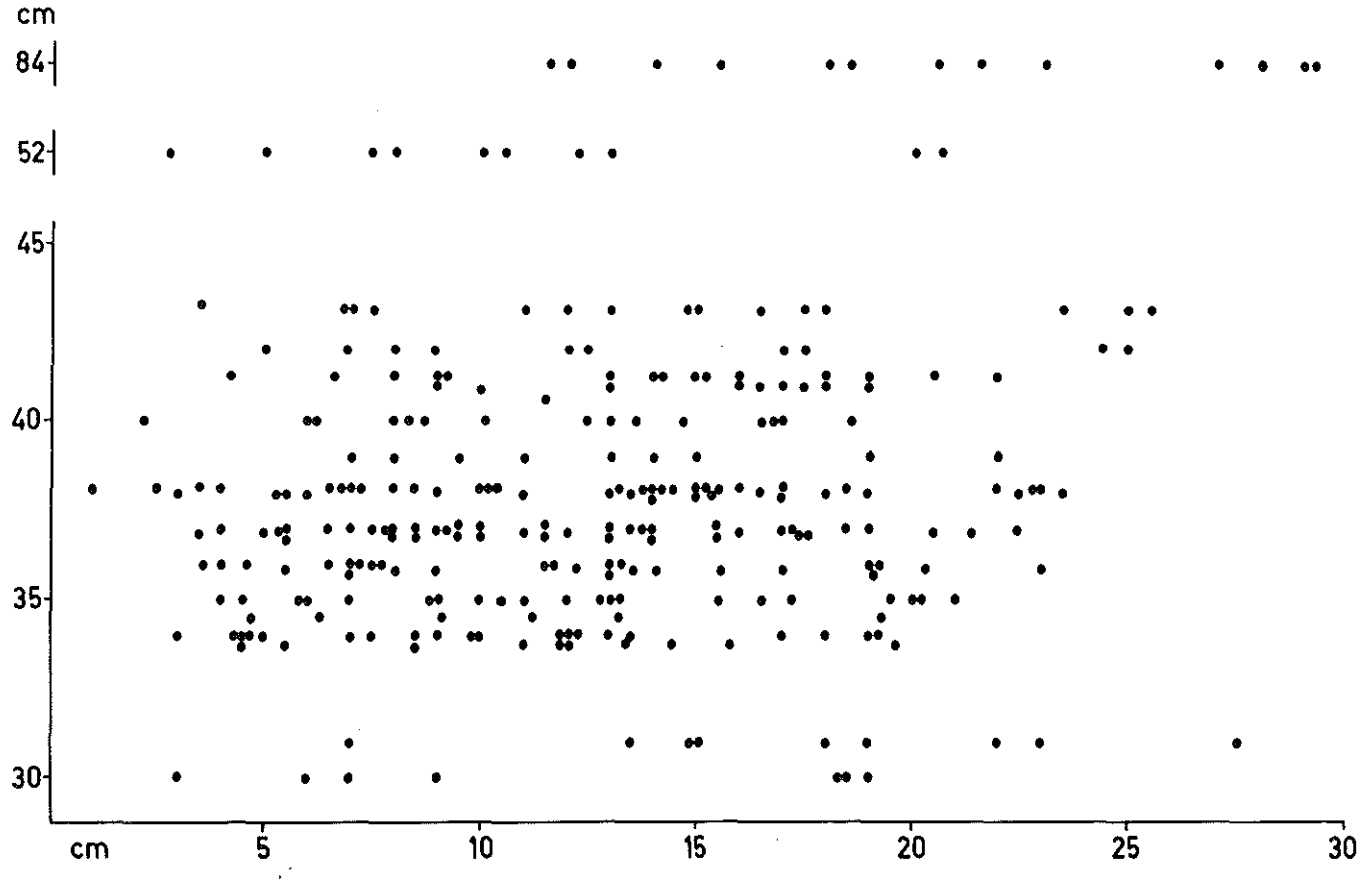


Table 2 Distribution of the stems of the ascending cerebral veins over the superior longitudinal sinus in 28 children with spina bifida aperta. The horizontal axis represents the distance from the torcular and the vertical axis the skull circumference.

Within individual specimens, the number of veins emptying into the superior longitudinal sinus was found to differ between the right and left sides. The location and distance from the torcular were also not symmetrical.

The number of ascending cerebral veins varied in the various specimens. On the right side a minimum of three to a maximum of eight veins were found, with an average of 5.3. On the left side the minimum was three and the maximum ten, the average 5.5. The smaller the number of entering veins, the larger the calibre.

In spina bifida aperta the number of stems of the ascending cerebral veins did not appear to be increased. Table 2 shows the distribution of these stems in 28 children with spina bifida aperta. None were found in the direct vicinity of the torcular. Frontally, the number diminished. There was an even distribution in the remaining part of the superior longitudinal sinus. Only in a child with extreme hydrocephalus (and spina bifida) were the stems of the ascending cerebral veins positioned at a large distance from the torcular. The distribution of the veins in moderate hydrocephalus gives the rough impression of an even expansion of the cerebral hemisphere in the early stages of hydrocephalus.

In several specimens the ascending cerebral veins changed direction close to the sinus, but sometimes only in the wall of the longitudinal sinus. In some cases this change of direction was so extreme that instead of emptying into the sinus in the frontal direction the vein turned sharply to enter the sinus in the direction of the torcular, or *vice versa* (Fig.17). The vein was also seen to divide close to or in the wall of the sinus, entering the sinus as two veins in some specimens. The course of the ascending cerebral veins were directed frontally on the right side in 63 per cent of the cases and toward the torcular in 37 per cent of the cases. On the left side these percentages were 58 and 42, respectively.

4. The other descending supratentorial veins and sinuses

The drainage of blood from the occipital lobes usually occurred via one to at most three veins going to the right and left transverse sinuses, only an occasional vein running from this lobe to the superior longitudinal sinus.

There were many anastomoses between the superficial veins.

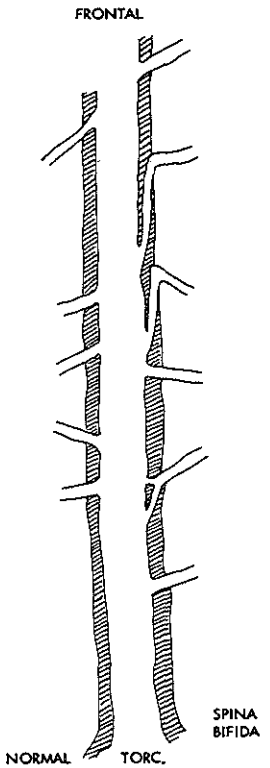


Fig.17 Drawing of the superior longitudinal sinus. At the left the distribution of joining cerebral veins from a normal specimen, at the right the distribution of these veins and the course near the sinus in a case with spina bifida aperta. Note the acute angle taken within the dural sheath.

The abundance of these anastomoses made it difficult to distinguish between cortical areas from which the blood was drained via descending veins.

The middle cerebral veins converged close to the origin of the sigmoid sinus, and proved to have an unusually wide calibre.

The petrosquamous sinus was easily recognized in all the specimens. It was not impressed in the skull at any point along its course, and ran more often on the internal than on the external surface of the dura. There were fine anastomoses with the cortical veins. The petrosquamous sinus and the middle cerebral veins sometimes formed a confluence at the origin of the sigmoid sinus. In these instances there was a large connection with the venous network on and around the sella into which the anterior cerebral veins and the striate veins drained and from which the basal veins arose.

5. a. Position of the torcular with regard to the foramen magnum

The ossification of the occipital bone around the foramen magnum was often irregular in children with spina bifida aperta. In many cases the posterior margin of the foramen consisted of fibrous tissue, as a result of which the bony foramen showed a local incisura. Although the measurements were done in specimens in which the bony foramen had not been dissected, it was clear that in spina bifida aperta patients the foramen magnum was often larger in the antero-posterior (depth) than in the transverse direction (width), the foramen in normal children being more round.

In both normal children and those with spina bifida aperta, the ratio between depth and width decreases as the size of the foramen magnum increases, but for equal width the depth was consistently greater in children with spina bifida aperta than in normal children. The difference increased with increasing size of the foramen magnum (Table 3).

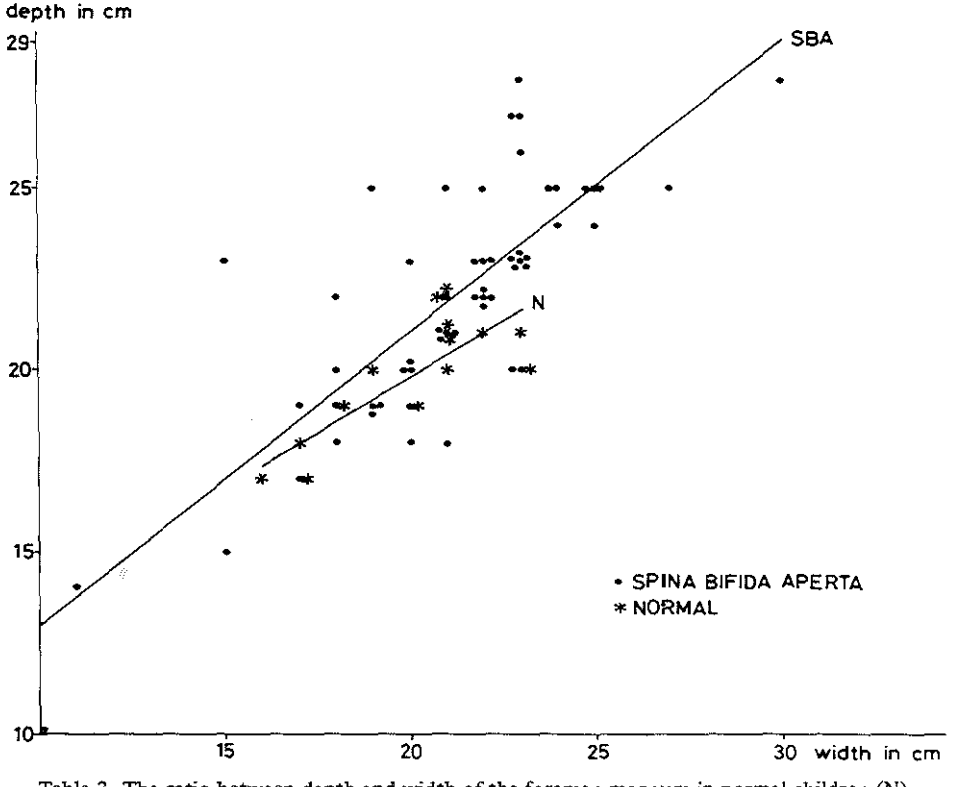


Table 3 The ratio between depth and width of the foramen magnum in normal children (N) and those with spina bifida aperta (SBA).

The measurements were classified according to age and assigned to three groups, group A comprising ages up to 9 days, group B those between 9 days and 9 weeks, and group C from 9 weeks to 9 months (Table 4 and 5).

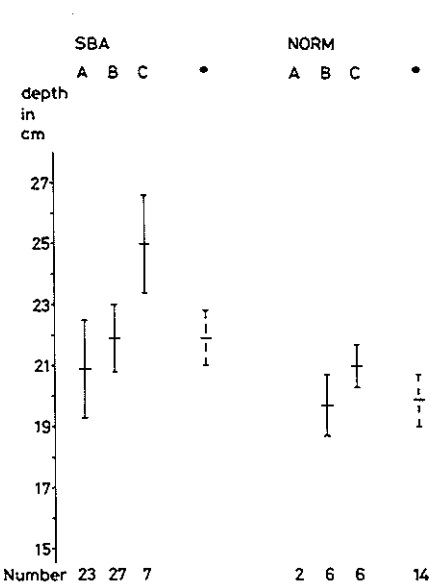


Table 4 The depth of the foramen magnum in children with spina bifida aperta and in normal children. The readings were grouped according to age (see text). The dotted line represents the mean with the age taken into consideration.

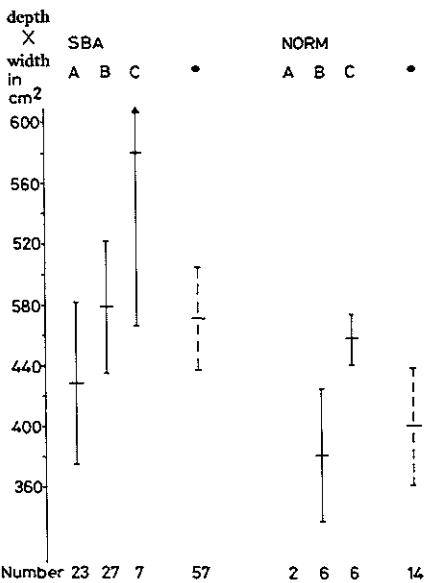


Table 5 The product of width and depth of the foramen magnum being a measure for the area of the foramen. The group of the normal children under the age of 9 days was too small to calculate a mean.

There was a positive correlation between age and both the depth and the product of the values for depth and width, the latter being a measure for the area of the foramen magnum. With the age taken into consideration, a significant difference ($p < 0.05$) existed between children with spina bifida aperta and normal children with respect to the depth and the area of the foramen magnum, both these values being larger in children with spina bifida aperta.

It is generally accepted that in the great majority of the cases the torcular lies closer to the margin of the foramen magnum in children with spina bifida aperta. To determine whether there is also

a difference in the distance from the torcular to the ventral margin of the foramen magnum, the findings in 27 specimens originating from children with spina bifida aperta were compared with those of 14 normal children. The results are shown in Table 6. As could be expected, the torcular also lay closer to the ventral margin of the foramen magnum in spina bifida aperta patients. This was statistically highly significant ($p < 0.01$).

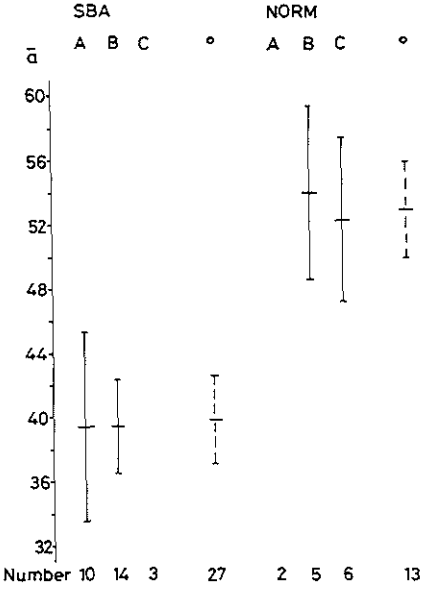


Table 6 The distance of the torcular to the ventral margin of the foramen magnum. The dotted line represents the mean regardless of the age.

An interesting feature was that no correlation with age was found. Apparently the position of the occipital bone changes during the growth of the foramen magnum, but in such a way that the distance from the torcular to the ventral margin of the foramen magnum remains the same.

The structure of the torcular Herophili clearly indicated its origin from the tentorial plexus. Its cavernous structure with many septa and connecting passages often made it difficult to determine the drainage pattern. The impression was obtained, however, that in children with spina bifida aperta the blood from the superior longitudinal sinus ran more often to the right transverse sinus and the blood from the straight sinus to the left transverse sinus. This is also the case in normal children (Padget 1957).

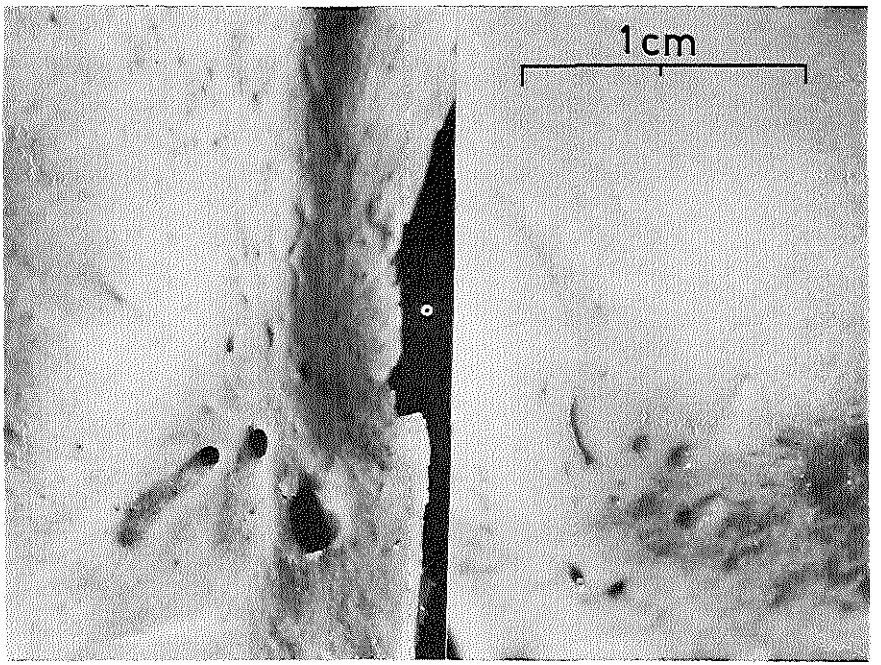


Fig.2 Two views of the occipital bone at the site of the torcular, at the left from the inside of the skull, at the right from the outside. One (in other cases several) vessel(s) from the torcular communicates with diploic veins, which run in a thickening in the midline of the occipital bone.

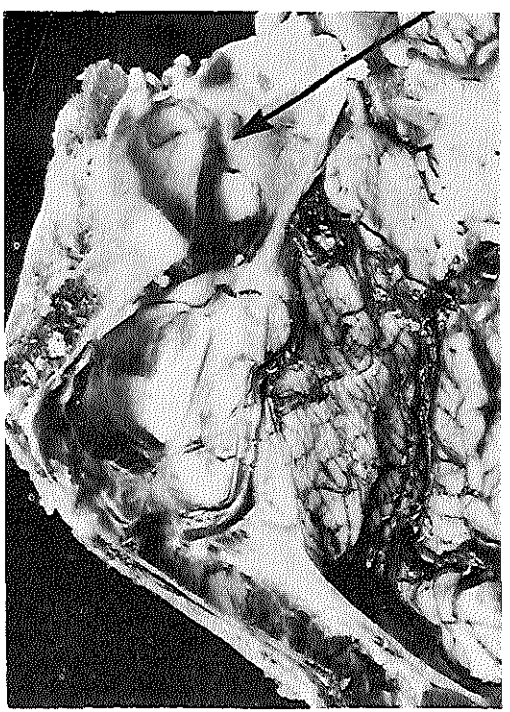
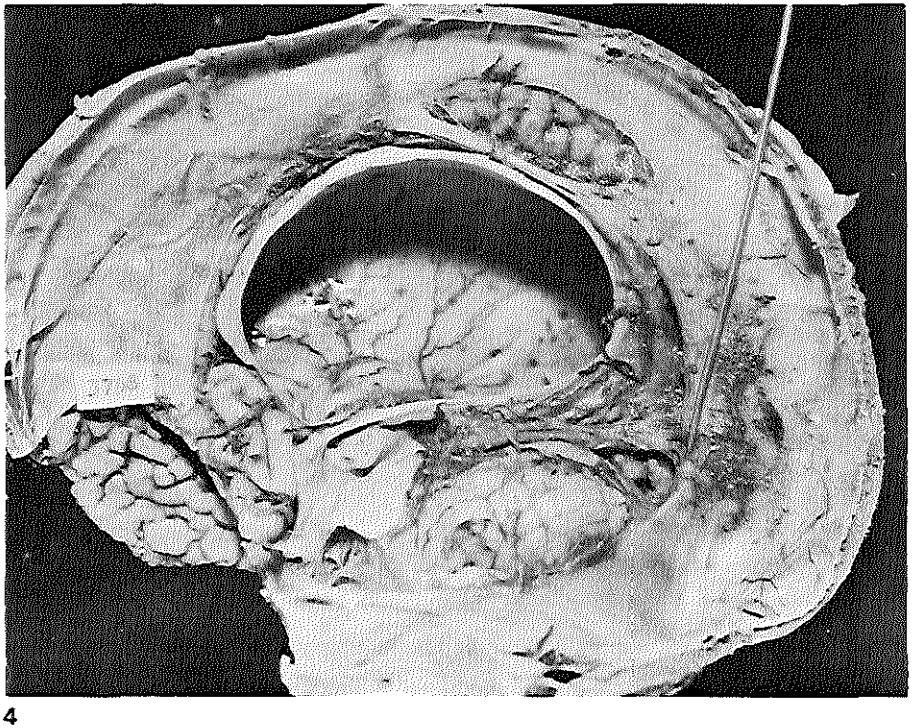


Fig.3 View from above after removal of the left cerebral hemisphere and supratentorial part of the skull. An emissary vein (arrow) runs through the foramen ovale and joins the superior petrosal sinus.



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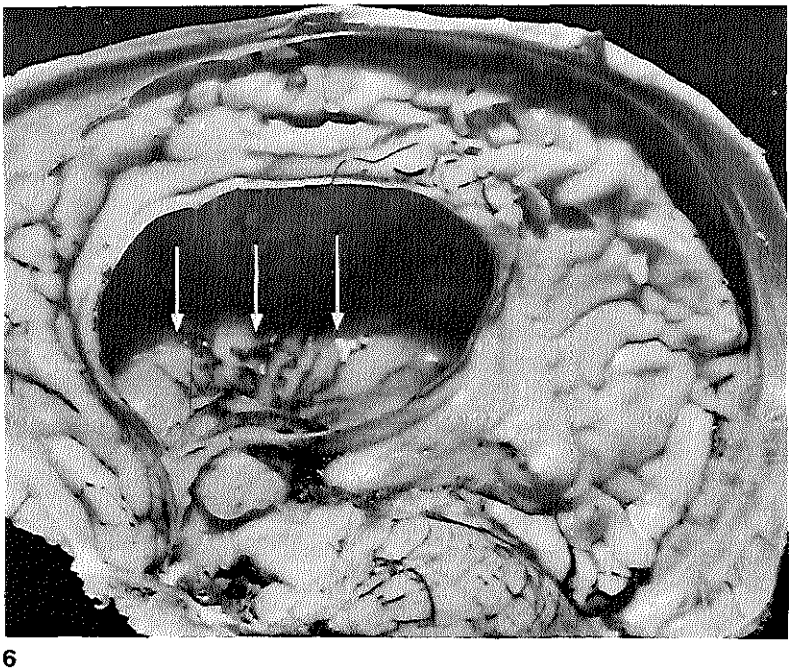

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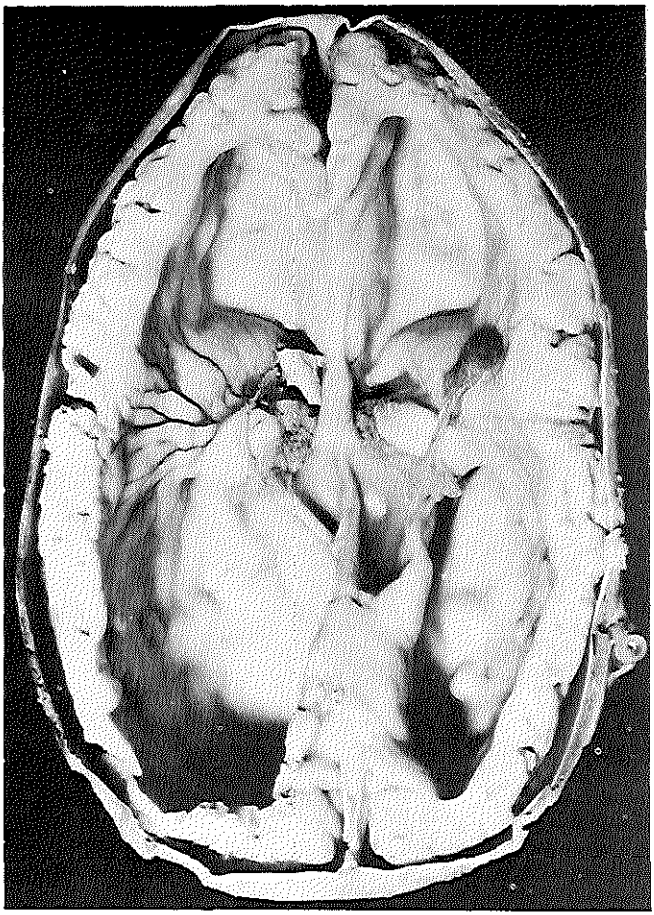
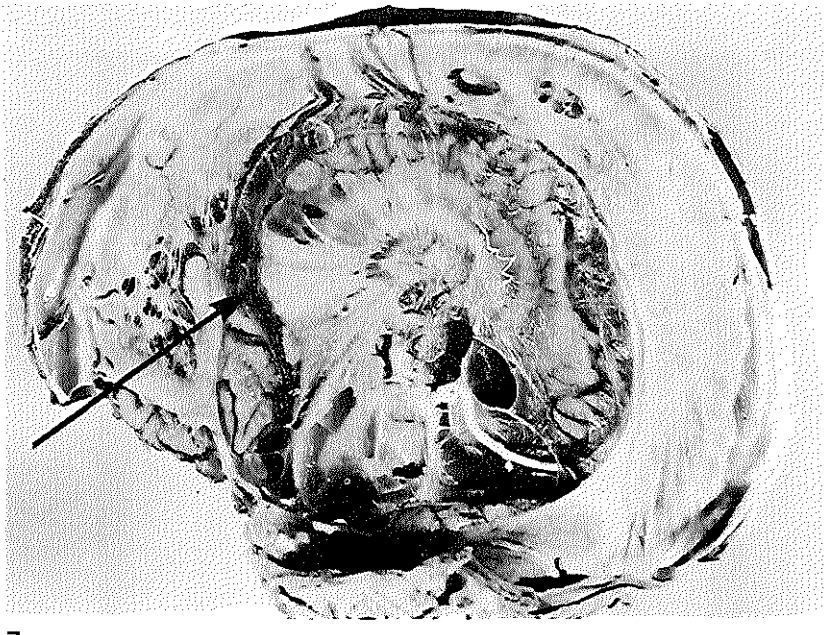


5

Fig.4 View of the medial aspects of the right cerebral hemisphere with the falx, tentorium and cerebellum *in situ*. Through the perforated septum pellucidum the subependymal tributaries to the thalamostriate vein can be seen. A probe has been passed through the sinus to show its position and direction.

Fig.5 View of the ventricular system from the vertex. The tributaries to the thalamostriate vein, which runs to the foramen of Monro, show clearly.

Fig.6 The veins running subependymally in the lateral ventricle (arrows) are important branches of the thalamostriate vein.



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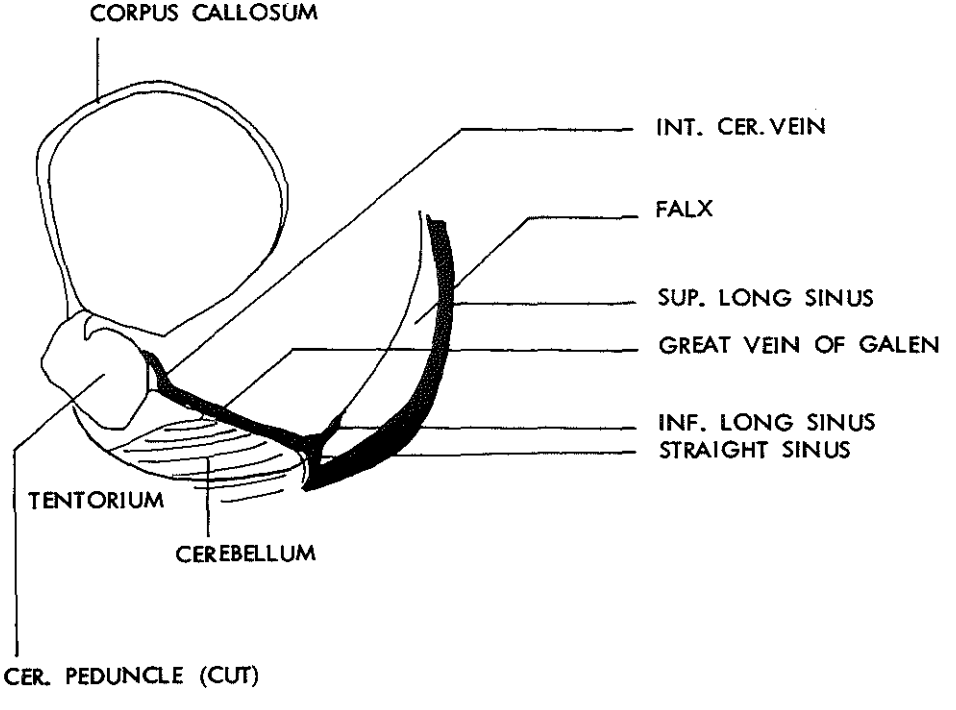
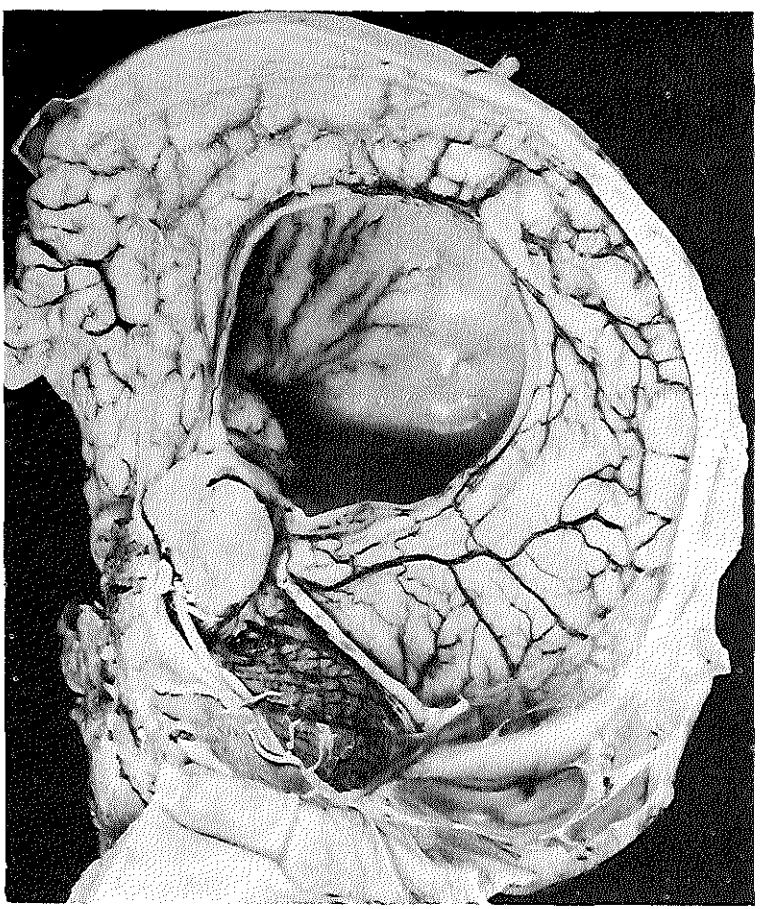

CORPUS CALLOSUM
INT. CER. VEIN
FALX
SUP. LONG SINUS
GREAT VEIN OF GALEN
INF. LONG SINUS
STRAIGHT SINUS
TENTORIUM
CEREBELLUM
CER. PEDUNCLE (CUT)

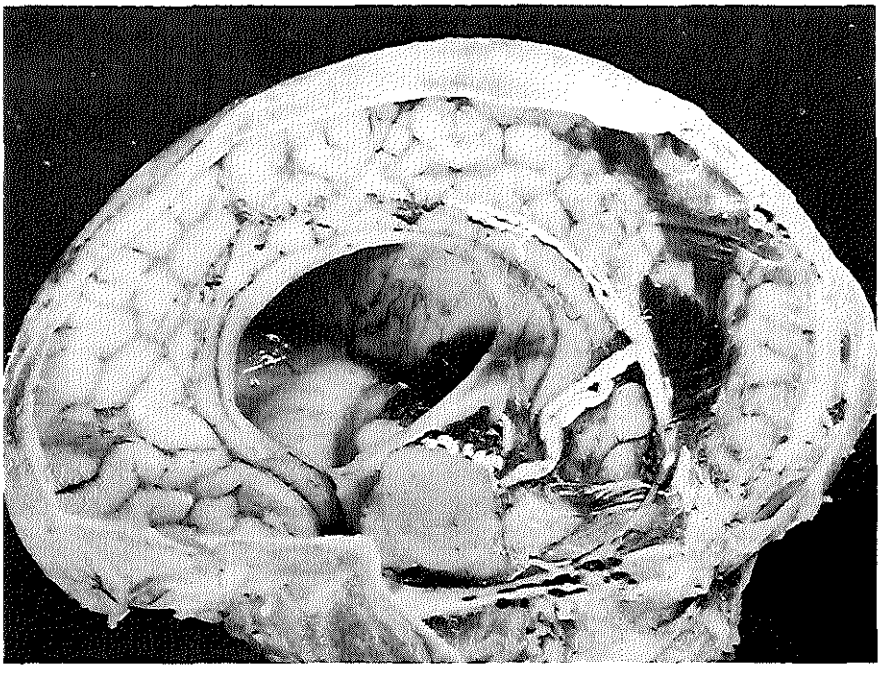
8a

Fig.7 In this specimen the great cerebral vein of Galen (white) is extremely long (12 cm). The anterior cerebral vein (not injected; arrow) communicates with the dural anastomoses in the falx at the site of the anterior fontanelle.

Figs.8[a] and [b] The great vein of Galen (white) runs over the cerebellum to the straight sinus. There is no inferior longitudinal sinus of any significance in this specimen.

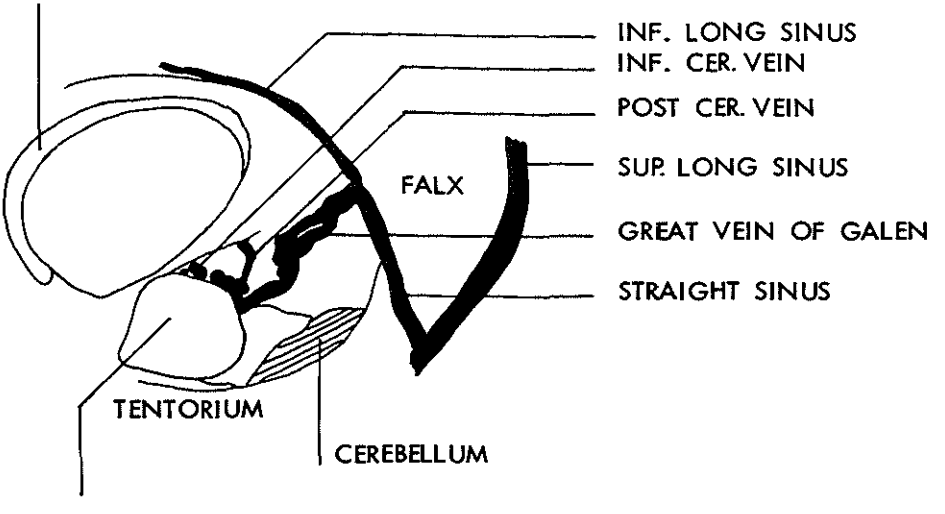


8b



9a

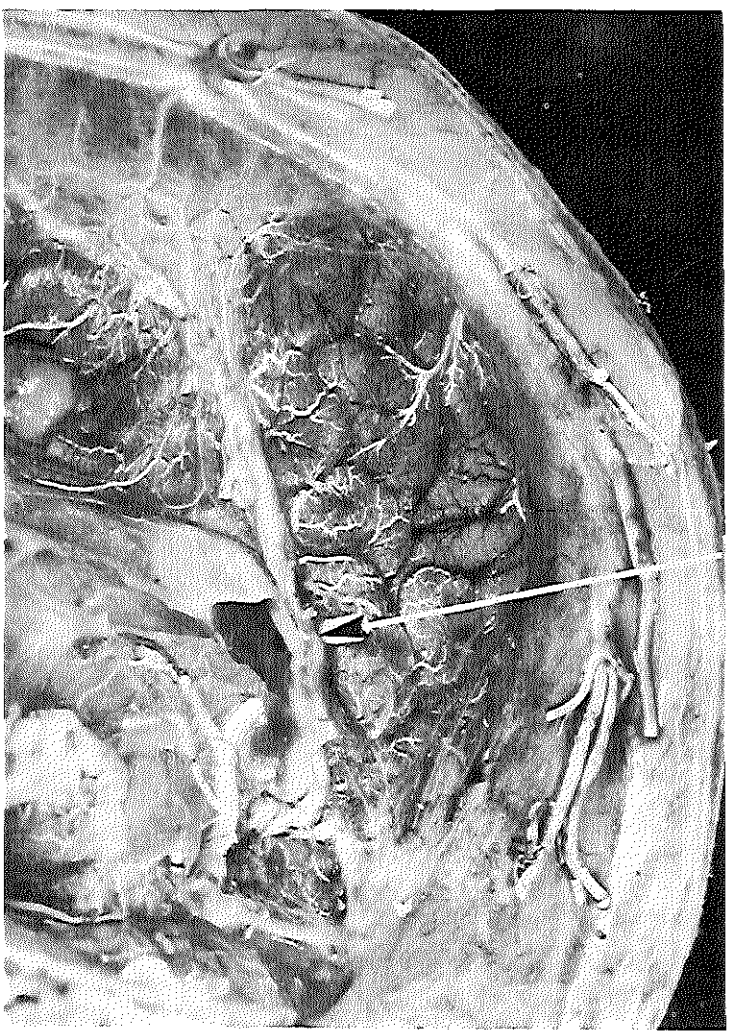
CORPUS CALLOSUM



9b

Figs.9[a] and [b] The great vein of Galen first ascends and then runs at an acute angle down to the torcular. In this specimen the angle lies in the tentorial notch. The vein of Galen is partially duplicated.

Fig.10 The falx is fenestrated in its middle third. The inferior longitudinal sinus runs in a narrow band of dura (arrow). Note also obliquity of the veins running to the longitudinal sinus at the posterior side.



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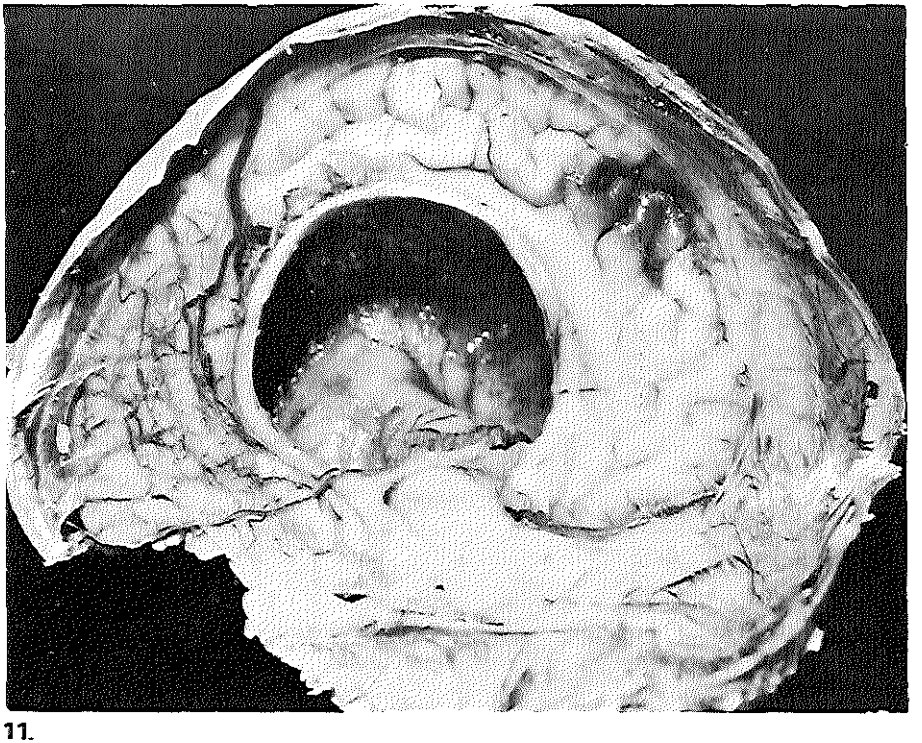

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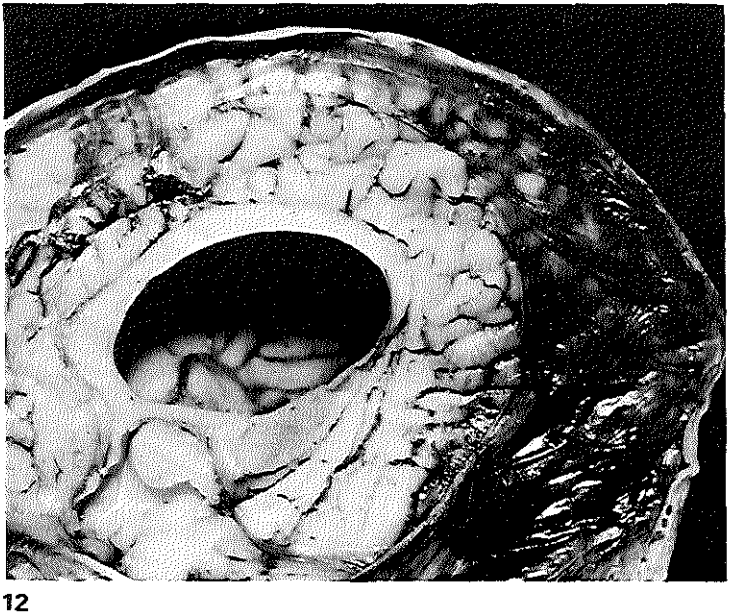

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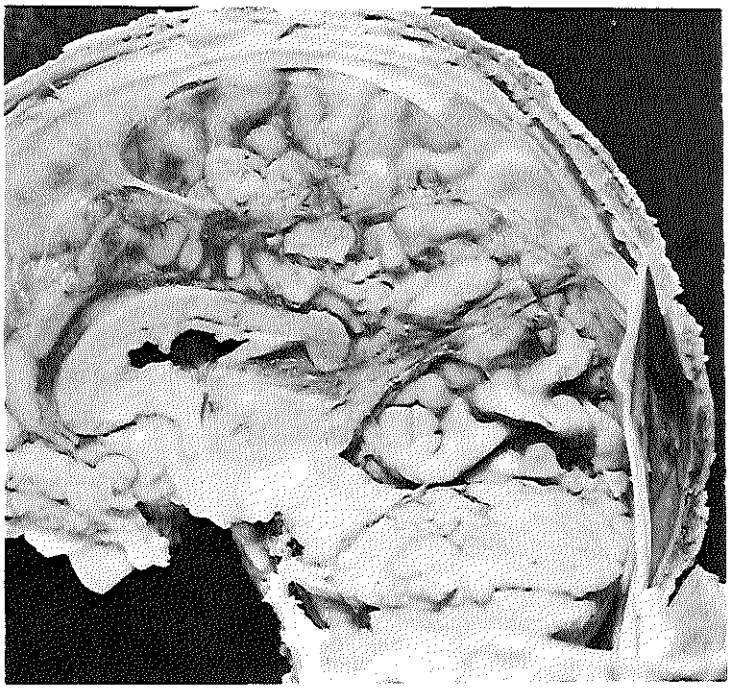
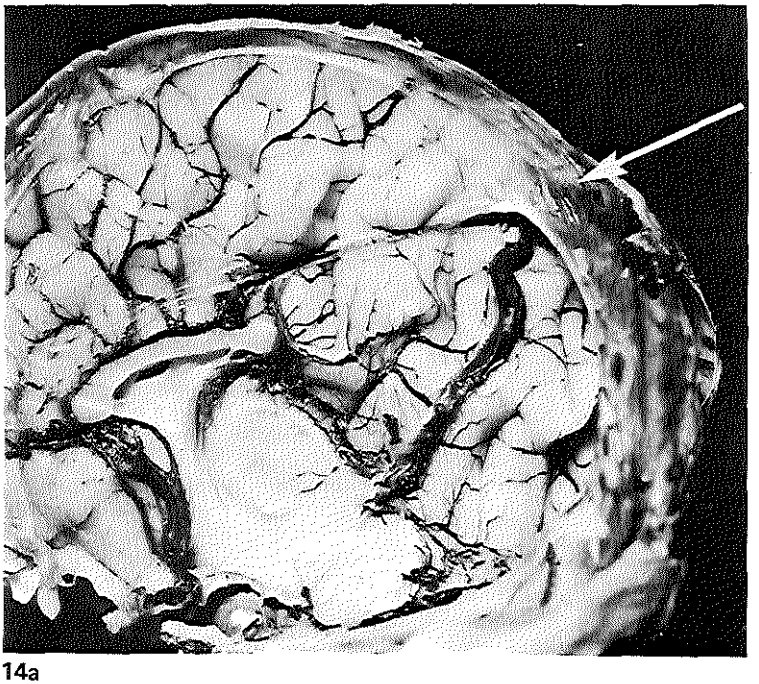


13

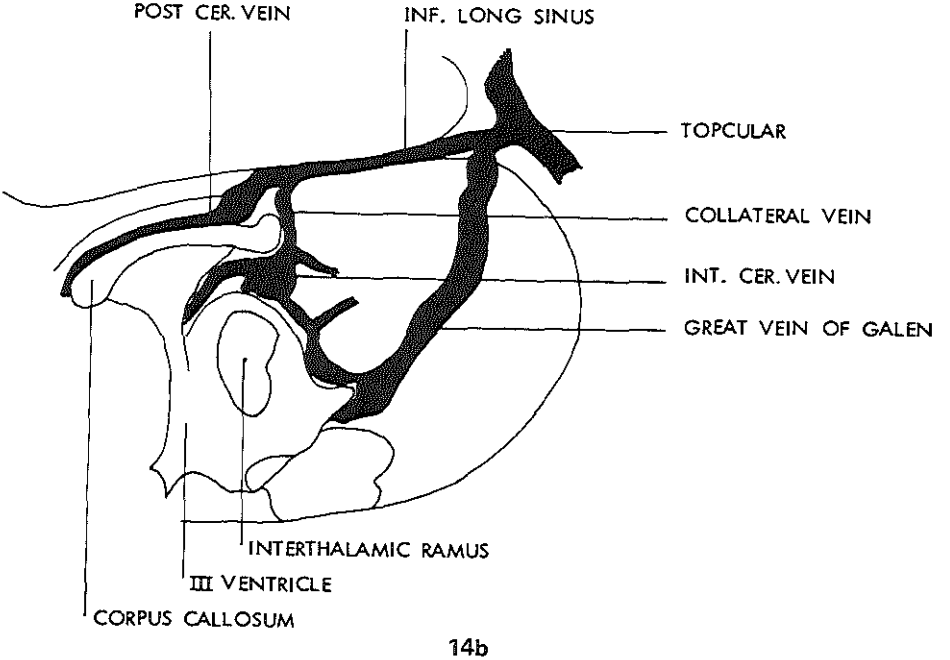
Fig.11 A distinct sinus runs transversely through the falx cerebri to the superior longitudinal sinus in the free edge of the falx, which shows a defect in its posterior two-thirds of its length. Branches from the anterior cerebral vein drain into this sinus.

Fig.12 Underneath the anterior fontanelle there are several sinuses running transversely through the falx cerebri to the superior longitudinal sinus.

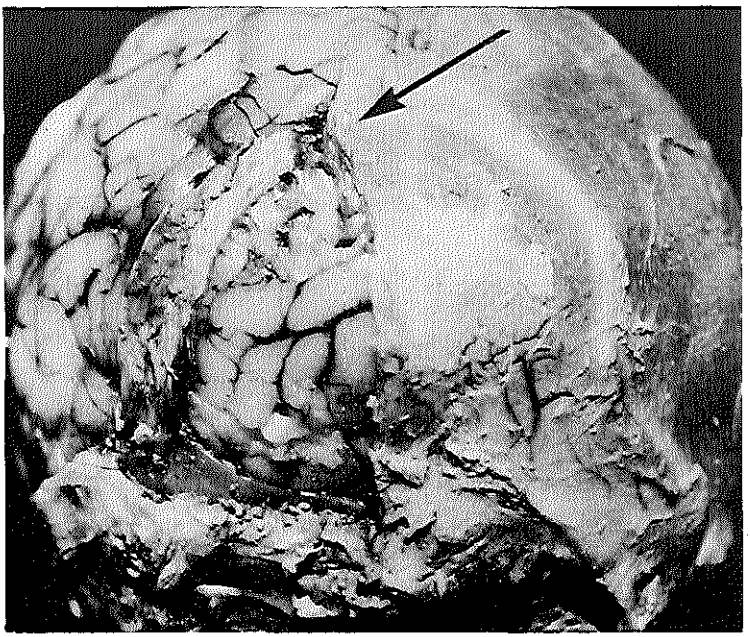
Fig.13 The great cerebral vein of Galen drains directly into the superior longitudinal sinus high above the torcular. There is no tentorium.



14a


POST CER. VEIN
INF. LONG SINUS
TOPCULAR
COLLATERAL VEIN
INT. CER. VEIN
GREAT VEIN OF GALEN
INTERTHALAMIC RAMUS
III VENTRICLE
CORPUS CALLOSUM

14b



15

Figs.14[a,b] and 15 The torcular (arrow) can be seen at the junction of the lambda sutures, with the vein of Galen ascending to it.

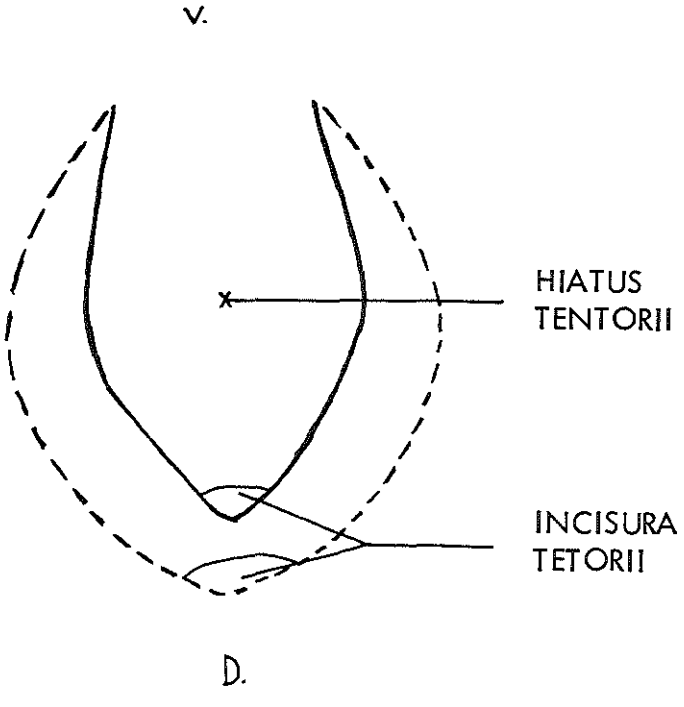


Fig.18 The hiatus tentorii and incisura tentorii normally (solid line) and in spina bifida aperta (dotted line).

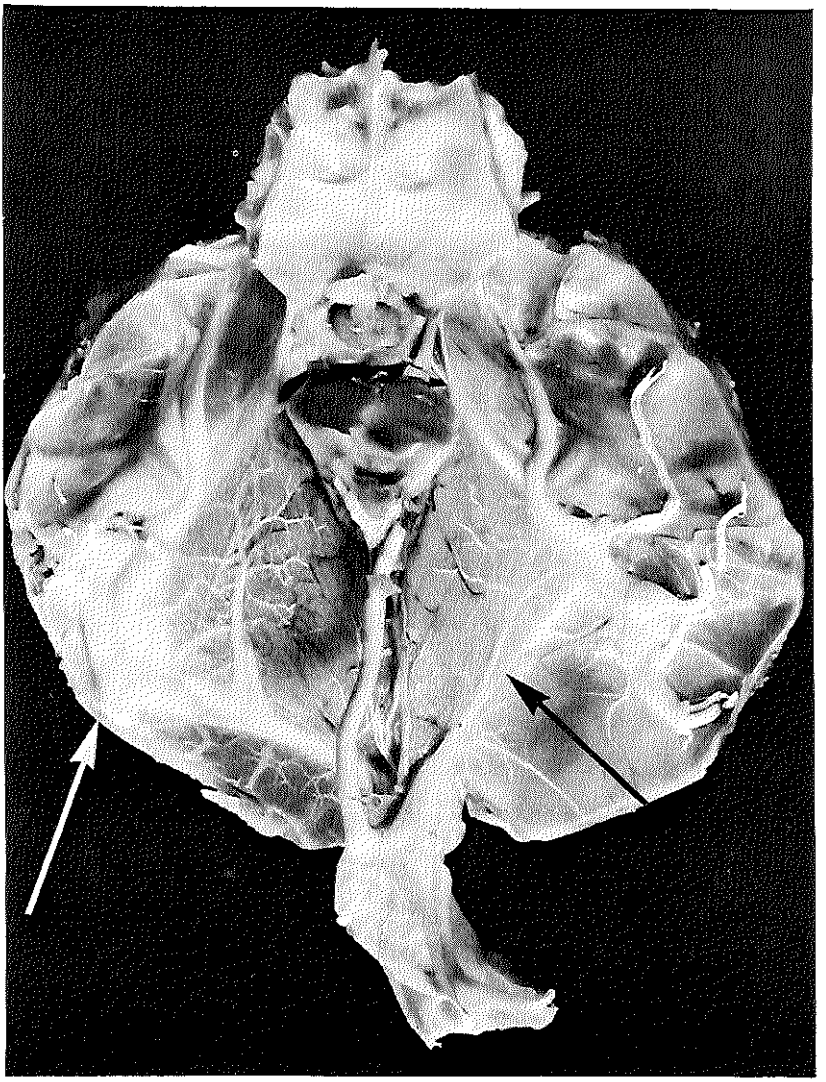


Fig.19 View from above after removal of the cerebral hemispheres and supratentorial part of the skull. The upper part of the cerebellum is drained by veins emptying into sinuses in the tentorial leaves and running to the lateral sinuses. There is an important confluence on both sides at the origin of the sigmoid sinus (white arrow). The right lateral sinus is stenotic (black arrow).

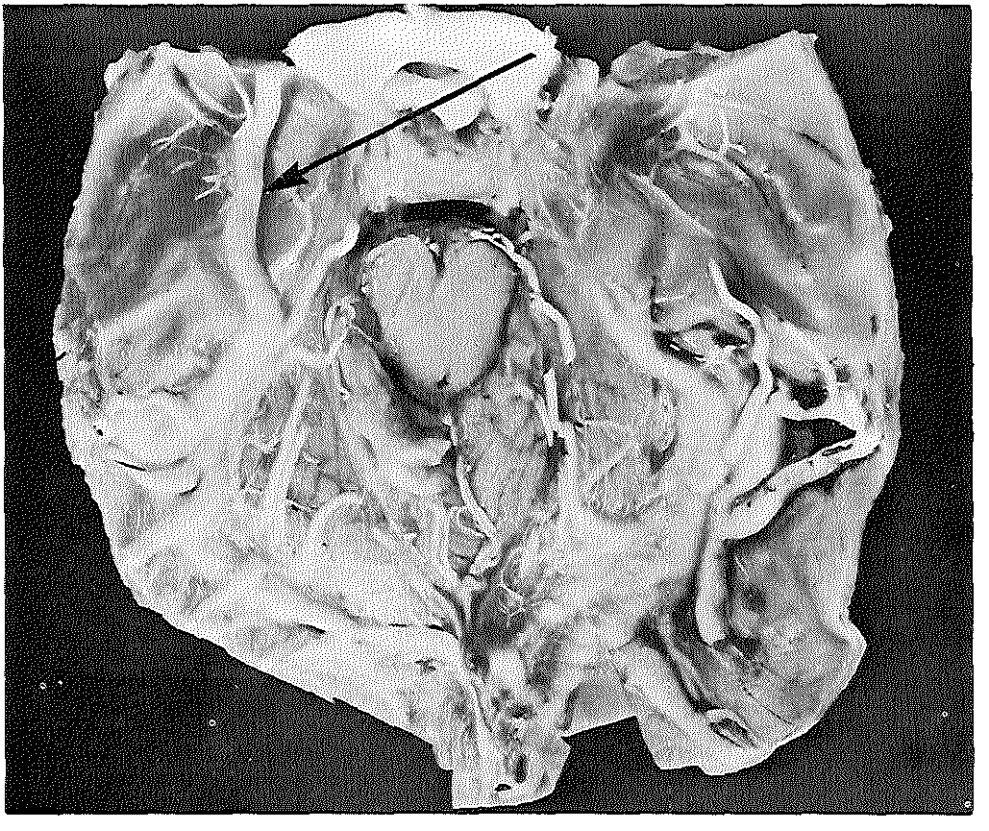


Fig.20 View from above after removal of the cerebral hemispheres and supratentorial part of the skull. In this specimen only one embryonic tentorial sinus (arrow) runs to the sigmoid sinus.

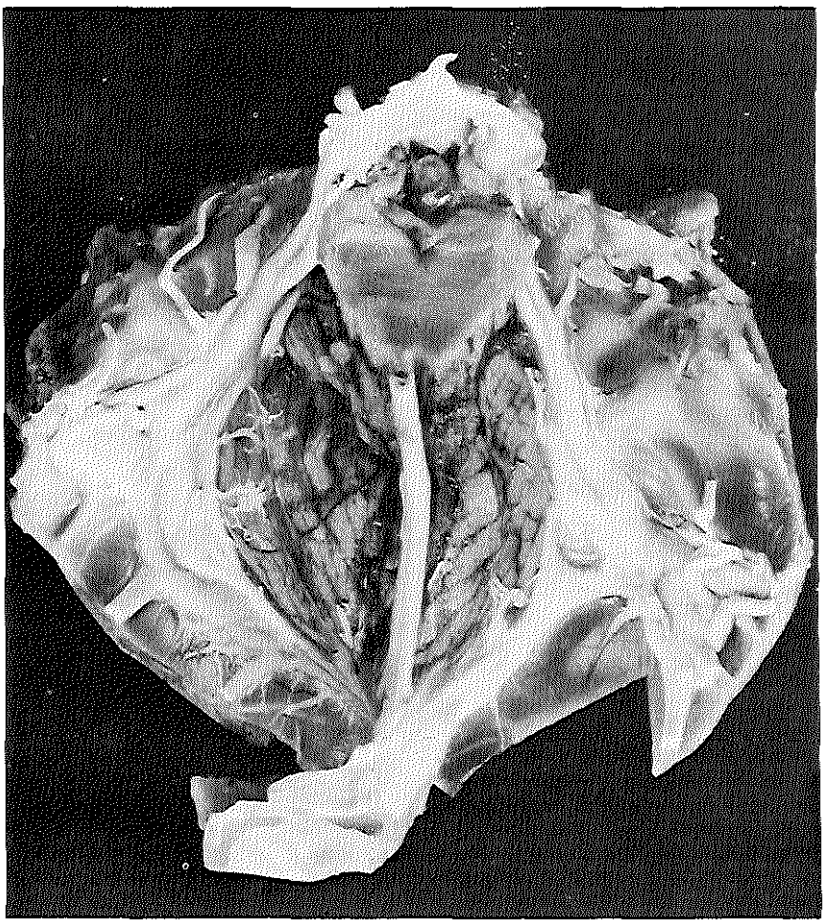


Fig.21 View from above after removal of the cerebral hemispheres and supratentorial part of the skull. Large veins and sinuses run to a confluence at the origin of the sigmoid sinus. Note thin tentorium.

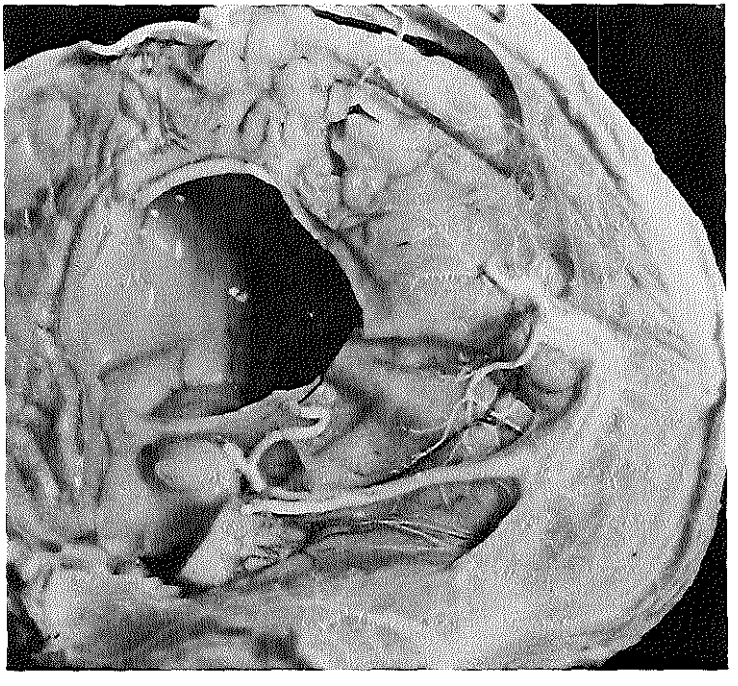


Fig.22 A duplicated inferior longitudinal sinus drains the medial aspects of the posterior part of the cerebral hemispheres. In the middle third of the falx such vessels are absent, causing a defect in the falx.

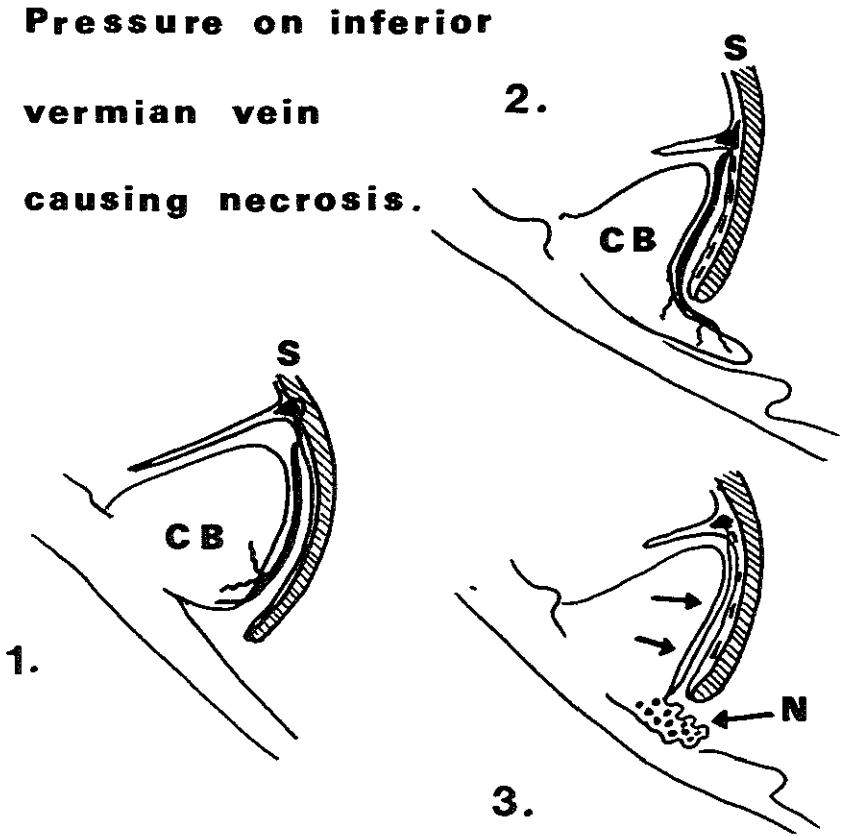


Fig.24 Impediment of the flow through the inferior vermian vein draining the cerebellar tonsils, which form part of the Arnold-Chiari malformation, in spina bifida aperta during intracranial hypertension causes haemorrhagic infarction and subsequent necrosis.

5. b. Veins and sinuses in the tentorium and over the middle fossa

a. The tentorium and the straight sinus

The tentorium was thin and narrow in all the specimens, and, like the falx, translucent. As a result of its abnormal position and the hypoplasia, the tentorial incisura was wide (Fig.18). The tentorium covered only a small portion of the cerebellum. The infratentorial space was small, but a large part of the cerebellum and mesencephalon lay above the plane of the tentorial hiatus. The upper surface of the cerebellar hemispheres was drained by veins converging to form one or more sinuses running through the tentorium to the transverse sinus (Fig.19). In specimens in which the tentorium could be clearly distinguished, these findings were made consistently (96 per cent of the cases). These sinuses are to be distinguished from the embryological tentorial sinus, which will be discussed next.

b. The tentorial sinus

A wide tentorial sinus frequently persisted in these children, receiving blood from the insular cortex and from deep cortical areas between the temporal and parietal lobes. Blood was also supplied by the anterior cerebral vein, and anastomoses with the middle cerebral veins were observed on the margin of the sphenoid. The sinus ran over the middle fossa in the posterior direction, passed medially around the eminentia arcuata, and then turned in a lateral direction, after which it ran parallel with the superficial petrosal sinus but did not unite with it. These sinuses drained into a confluence at the origin of the sigmoid sinus. The tentorial sinus occurred on only one side in some specimens (Fig.20).

c. A "confluence" at the origin of the sigmoid sinus

A confluence of this type was present in all the specimens. It was formed by the petrosquamous sinus, superficial middle cerebral veins, and a frequently present vein of Labbé, and, as already mentioned, received the superficial petrosal sinus and tentorial sinus (Figs.19 and 21).

d. The communications between the plexus around the sella with the emissaries

These communications lie extradurally. A few specimens showed

a vein draining the tip of the temporal lobe, from which it ran over the floor of the middle fossa and emptied into the confluence at the origin of the sigmoid sinus.

5. c. Drainage from (A) and via (B) the posterior fossa

A. In the normal situation the blood from the cerebral structures in the posterior fossa is drained via three systems. This arrangement could be clearly distinguished in children with spina bifida aperta.

a. The precentral cerebellar vein was easily distinguished in this series. The right and left branches usually united and invariably passed over the cerebellum, in direct contact with the latter, and emptied into the part of the straight sinus touching the cerebellum, thus running a course independent of that taken by the great vein of Galen or straight sinus. The insertion was thus not always on or near the transition between the vein of Galen and the straight sinus. The precentral cerebellar vein was accompanied by smaller veins which could be identified as the posterior mesencephalic vein and the superior vermian vein.

b. The inferior vermian vein joined the straight sinus close to the torcular. It took part in the displacement of the cerebellar tonsils into the cervical spinal canal and thus drained partly the Arnold-Chiari malformation. It ran in the median cerebellar fissure to the straight sinus. Numerous veins and sinuses running through the tentorium to the lateral sinuses (Fig.19) drained the upper part of the cerebellar hemispheres.

c. In our specimens the petrosal vein was easily distinguished, but its termination was often located so far laterally that it emptied into the confluence on the sigmoid sinus, as described above. No shunts with the basal veins were found in this series.

B. The closeness of the torcular to the foramen magnum in children with spina bifida aperta leaves little space for a normally differentiated sinus system in the posterior fossa. Adjacent to and posterior to the foramen magnum, the narrow dura had almost

entirely the structure of a plexus and in extreme cases formed a cavernous system in which no distinction could be made between the torcular, occipital sinus, and marginal sinus (Fig.23). This occipital plexus had a distinct communication with the vertebral plexuses. The sinuses in spina bifida aperta differed from those found in normal children in that they were not composed of triangular tubes with rigid walls running in grooves in the skull. These walls appeared weak and could be easily compressed by mechanical pressure applied to the wall.

The noteworthy deviations treated in this section concern the closeness of the torcular to the foramen magnum. The lateral sinuses ran near to the foramen magnum to the sigmoid sinuses. At the origin of the sigmoid sinus there was a confluence receiving the petrosal veins and veins and sinuses from the floor of the middle fossa and from the cerebral hemispheres. The embryonic tentorial sinus usually persists in these children. The inferior vermian vein draining the Arnold-Chiari malformation had close connections with the margin of the foramen magnum. The petrosal vein drained more laterally. The dura of the posterior fossa consisted of a plexiform structure in which no clear distinction could be made between the various sinus systems normally present in the dural layers of the posterior fossa, thus constituting an occipital plexus. The walls of the sinuses were weak and were vulnerable to mechanical pressure.

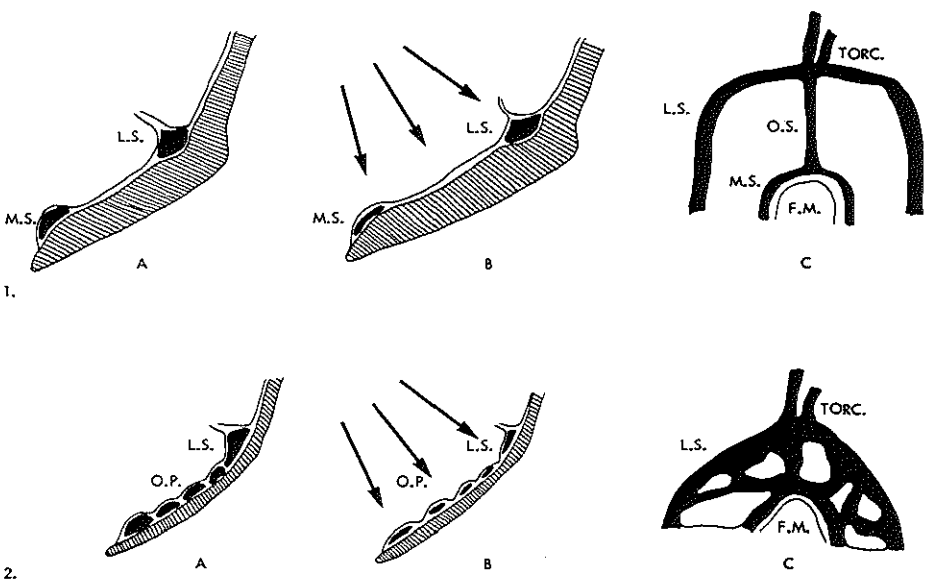


Fig.23 Diagram representing the dural venous system in the normal posterior fossa (1) and in spina bifida (2) aperta. 1B and 2B illustrate the changes during intracranial hypertension. The vessels of the dural system in spina bifida, which form an occipital plexus (O.P.), are more easily compressed because of their structure.

CHAPTER VI

DISCUSSION

a. Venous factors and spina bifida aperta

The cause of hydrocephalus in children with spina bifida aperta and in some of the cases of congenital hydrocephalus is sought in a malformation of the normal CSF pathways. Drainage via the spinal canal in children with spina bifida aperta may possibly prevent the formation of normal extra-cerebral pathways. Elevated ventricular pressure can also exist *in utero*, resulting in dilatation of the ventricular system in the developing brain.

Distinct anomalies of the intracanial venous system were found particularly in children born with spina bifida aperta, of whom about 80 per cent develop hydrocephalus. This raises the question of whether there is a causal relationship between hydrocephalus and the abnormalities of the venous system.

As pointed out in Chapter II elevated intracranial venous pressure may play a causal role in the development of hydrocephalus. The anatomical divergence of the cranial venous system in children with spina bifida aperta increases the chance of elevated intracranial venous pressure.

Anatomical venous anomalies occur consistently in the brain of children with spina bifida aperta. We are led to ask whether fetal hydrocephalus causes these variations or does the fetal spina bifida aperta brain develop according to a different pattern? It is difficult to decide which is the case. There are gross abnormalities in the brain in spina bifida aperta, even in children without hydrocephalus, so it

is conveivable that different inductions exist in the fetal development of this organ. Further study of the anatomy of the brain in spina bifida aperta is needed before we can determine which abnormalities are caused by hydrocephalus and which are primary.

In all of these children the transverse sinuses had an abnormal position and structure. The tentorium and the transverse sinuses were coalescent with the occipital and marginal sinuses and created an occipital plexus covering the posterior fossa. The transverse sinus was not positioned in a triangular dural structure as in the normal situation. Norell *et al.* (see page 24) have shown that in these children the transverse sinuses may be compressed, thus impairing the outflow of blood from the skull via the normal route. Fig.23 shows how increased pressure exerted on the wall of the posterior fossa may result in diminished blood flow through these channels. As a consequence alternative drainage routes become important.

In the midline these alternative channels are in the first place the dural anastomoses in the falx, at the site of the anterior fontanelle. These anastomoses connect the deep venous system with the superior longitudinal sinus and explain the increased drainage through the parietal foramen by the parietal emissary veins, which empty into the external jugular system. These dural anastomoses were found 40 times in 52 specimens.

The formation of the superior longitudinal sinus from the primitive marginal sinuses by unequal or alternate enlargement of the channel of one side, as well as by the coalescence of the bilateral channels, is illustrated by the duplications and partitions of the longitudinal sinus found in spina bifida aperta (Fig.17).

The fetal telencephalon expands backward in the formation of the occipital and temporal lobes. This expansion seems to occur when drainage patterns are fixed. In spina bifida aperta the stems of joining ascending cerebral veins frequently made an acute angle inside the dural sheath covering the sinus wall, as pointed out above (Fig.20). The stems can make an acute angle in the normal adult too (Stopford 1930), but in children this seldom occurs. In the few normal brains examined in the course of this study, an acute angle was never found. Investigation of larger series of normal brain is needed to clarify the configuration. The frontal direction taken by some of the ascending cerebral veins may be explained by the backward expansion of the fetal hemispheres, but the acute angle inside the dural

sheath seems to have been formed at a later date. Since the backward expansion occurs at a time when the dural sheath is till histologically immature, the ill-defined tributaries, representing remnants of the anterior dural plexus on the dorso-caudal convexity of the hemisphere, can easily change direction. The acute angle may very well have occurred at a time when the pathological backward expansion of the spina bifida aperta brain pushed the tentorium in a very low position. This situation may be aggravated by the simultaneous occurrence of internal hydrocephalus.

In three specimens showing marked ventricular dilatation the ascending cerebral veins took an extremely frontal direction (Fig.10); this might also be explained by the abnormal skull growth, which is of course most pronounced at the level of the vertex. Over the convexity of the cerebrum the ascending cerebral veins showed anastomoses with descending veins.

Because the autopsy method involved removal of the frontal part of the sinus, accurate measurements of the superior longitudinal sinus were not feasible. No measurements of its length in normals were available either.

During embryonic development the scalp region is a-vascular until the 20 to 40 mm stage. The emerging scalp plexus advances dorsally from all sides up over the vault of the developing skull toward the region of the future anterior fontanelle. At this site the plexus becomes anastomosed to the sagittal plexus, from which arises the superior longitudinal (sagittal) sinus. Emissaries at the site of the anterior fontanelle are therefore formed later than the basal emissaries, which are present in the chondrocranium (Padget 1957). Hence, we are not surprised that in the brain in spina bifida aperta the formation and persistence of collaterals in the falx has taken place underneath the anterior fontanelle. In children with spina bifida aperta the scalp region overlying the anterior fontanelle often shows a venous plexus from which draining veins run in all directions to the face and neck.

Another finding was that the presence of transverse collaterals anteriorly in the falx caused the formation (or the persistence) of a distinct falx structure, while posteriorly where no sinuses ran in the mesenchymal tissue of the great longitudinal fissure, the falx often failed. Thus, there seems to be a relationship between the formation of the falx and the presence of sinuses in the great longitudinal

fissure.

Pronounced sinuses in the tentorium may be important as collateral channels when compression of the transverse sinus occurs. A frequently present confluence at the origin of the sigmoid sinus indicates that the drainage via the torcular may be less important.

The large veins which leave the skull in the middle fossa show other connections with the venous plexuses of the face.

An important tentorial sinus always persists in these children after birth, and provides for drainage of blood from deep central brain areas and from the veins of the Sylvian fissure. Drainage along the spinal column also seems to be pronounced.

Some of the variations may be related to an adaptation to the abnormal structure, resulting in enhancement of the drainage of blood. In some instances this relationship is less clear, particularly in the case of the great vein of Galen and the straight sinus. These variations might be the consequence of the development of the hydrocephalic brain with divergent inductions.

The dilatation occurs mainly in the lateral ventricles and to a lesser degree in the third ventricle; in the fourth ventricle it is rare. The increased skull growth is more pronounced at the vertex than at the base, and this seems to be the main reason for the unequal ventricular dilatation. The pressure on the skull base gives typical variations, however.

Gardner (1965) thought that the relatively great increase of the cerebral volume supratentorially was the cause of the abnormal situation of the tentorium and the transverse sinuses, but this does not explain the two courses of the great vein of Galen and straight sinus. The present investigation did not clarify this problem.

The investigations of the intracranial pressures by Becht, Weed and Flexner, Wright and Bedford were all performed in dogs, where the CSF pressure did not influence the intracranial venous pressure in the dural venous system under physiological conditions. However, a rise in the dural sinus pressure caused a rise in the CSF pressure. These findings are in all probability applicable to the human.

It must be kept in mind that such pressure relations could hardly exist in the intracerebral veins and in the veins running in the subarachnoid space, if they did there would be no conceivable way for the blood to leave the brain. The pressure in these veins must be equal to or slightly higher than the CSF pressure. In the normal brain

the venous pressure would then be dependent on the CSF pressure, whereas different basic principles determine the pressure in the dural sinus system.

In children with untreated hydrocephalus and spina bifida aperta, in children with hydrocephalus from other causes, and in experimental hydrocephalus the dural venous pressure is elevated. In addition, the dural venous pressure in children with spina bifida aperta has a parallel relationship with CSF pressure, a phenomenon which does not exist in other forms of hydrocephalus. Then the pressure in the sinus system equals the pressure in the cerebral veins, and in its physiological behaviour the sinus system could be considered to function in these children as subarachnoid or intracerebral veins.

This situation undoubtedly influences the drainage of blood from the brain in children with spina bifida aperta, and has repercussions on all the intracranial pressures. After each systole, a certain amount of blood is forced into the cranial cavity, which would result in a volume increase at the cost of a dangerous intracranial pressure rise unless an equal volume left the skull. This volume is made up of the combined amounts of CSF and venous blood leaving the skull during each systolic rise of the blood pressure. CSF is transported to the spinal canal, where space can be created for it when blood is squeezed from the peridural venous plexus. Secondly, blood flows into the sinus system and out of the skull, which is perhaps the most important factor here.

This damping effect, is diminished during increased CSF pressure in children with spina bifida aperta. Passage of the CSF to the spinal canal is hampered by the presence of a Cleland or Arnold-Chiari malformation and is possibly negligible when the spina bifida is closed surgically. Furthermore, the drainage of venous blood is not optimal either. Although the elevated CSF pressure in the ventricular system is regarded as the primary factor for the ventricular dilatation, the pathological dural venous system combined with the Cleland or Arnold-Chiari malformation may also contribute to maintaining the elevated CSF pressure or even lead to a further increase. Apart from the static factor of continuous excessive pressure, we must take into account the detrimental effect on the brain of the systolic pressure wave, which here is not alleviated by an adequate damping effect. Possibly, however, in these children the systolic pressure wave is partially absorbed by the elastic skull with its wide

sutures and open fontanelles.

During increased intracranial pressure, venous stasis is more easily promoted in spina bifida aperta and haemorrhagic infarction may be the result in the cerebral hemispheres. Areas of cerebral necrosis are often found, particularly in the occipital lobes.

The first portion of the inferior vermian vein, draining the herniated cerebellar tonsils forming part of the Arnold-Chiari malformation, may be compressed on the dorsal margin of the foramen magnum during intracranial pressure rises. As the result of venous statis, haemorrhagic infarction and subsequent necrosis of the cerebellar tonsils may occur (Fig.24). On casual inspection in these cases an Arnold-Chiari malformation may seem to be absent in the presence of spina bifida aperta, but the deformity of the fourth ventricle and the caudad displacement of the medulla oblongata are nevertheless indicative of the previous presence of an Arnold-Chiari malformation in these cases.

b. Venous factors and arrest of hydrocephalus

The question remains whether adequate treatment of the hydrocephalus can improve the venous drainage of the brain and promote consequently the chance of arrest of the hydrocephalus without artificial drainages. De Lange (1968) suggested that improvement of the venous drainage of the hydrocephalic brain might facilitate the transfer of fluid and electrolytes from the brain parenchyma to the blood vessels and consequently from the ventricles to the brain, thus promoting arrest of the hydrocephalus.

There is no evidence that the resorption of CSF is normalized post-operatively and that normal CSF pathways develop when the hydrocephalus is adequately treated. Therefore, arrest is not promoted by the formation of a normal subarachnoid space and arachnoid villi. Hydrocephalic children with spina bifida aperta did not show consistent modification of the anatomic variations of the venous drainage. Consequently, if any alterations of the venous drainage were to occur, it would be more in a physiological than in an anatomical sence. Such changes could only be observed if investigations like those performed by Norrell *et al.* could be done in children with arrest of their hydrocephalus, but this cannot be considered ethically possible.

A well-functioning ventriculo-cardiac drain is essential in children with progressive hydrocephalus, undoubtedly because it provides for both the possibility of transport of CSF and the damping of the systolic pressure wave.

Long-standing decompression by a valve system gives a rostral shift of the brainstem (Emery 1956). Theoretically, the "ball-cock function" of the Cleland or Arnold-Chiari malformation and compression of the sinus system in the posterior fossa could then be abolished or diminished. If this occurs during the systolic pressure rise, displacement of CSF to the spinal canal and to the basal cisterns and outflow of venous blood from the cranial cavity may be promoted.

c. Conclusions

On the basis of the present results, we may conclude that increased CSF pressure in the cerebral ventricular system in children with spina bifida aperta can lead to increased cerebral venous pressure and dural sinus pressure. As a consequence, there is a greater disproportion among the intracranial fluid pressure relations. This vicious circle can be broken by adequate CSF drainage, while the abnormal cerebral venous system remains unchanged. Since the venous variations persist during further life, a recurrence of increased ventricular CSF pressure can have the same disastrous consequences. This too means that children who show an arrest of hydrocephalus and who have no functioning CSF shunting device, must be checked regularly and thoroughly.

The formation of alternative pathways for the venous drainage of the brain implies that in some of the cases of spina bifida aperta intracranial venous hypertension, or at least impairment of the venous outflow, develops early in fetal life. In the cases in our series without hydrocephalus the venous system was also highly abnormal. This supports the view that the changes in the venous system are only partially due to the hydrocephalus. They probably form part of the developmental malformation of the brain in spina bifida, of which the hydrocephalus is but one manifestation.

Our findings with regard to the falx cerebri are very similar to those of Peach (1956), but he did not discuss the veins in his paper.

The obliteration of the great longitudinal fissure, reported earlier by Cameron (1957), seemed to be caused by the small, hypoplastic, and incomplete falx.

The abnormalities of the falx require further study. In cases of congenital hydrocephalus not attented by spina bifida the falx is a firm structure showing a normal configuration. The atrophic falx in spina bifida is not interpreted as pressure atrophy then. It is an intriguing problem why the mesenchymal structures surrounding the brain in spina bifida aperta are deficiently formed.

In children with spina bifida aperta the external jugular system is essential for the venous drainage of the brain during increased CSF pressure. This explains why the skull veins are engorged with blood in these children with intracranial hypertension, and also clarifies the dilatation of the common facial vein.

SUMMARY

The intracranial venous system was investigated macroscopically in children with spina bifida aperta. Attention was drawn to this system by the marked dilatation shown by the scalp veins in cases of progressive hydrocephalus. Furthermore, during the application of a ventriculo-cardiac drainage for the treatment of the hydrocephalus, the common facial vein was often found to have an extremely wide calibre.

Initially specimens from hydrocephalic children were investigated without regard to the cause of the hydrocephalus, but after noting that severe anomalies occurred regularly in children with spina bifida aperta, we limited the study to these cases. This limitation had the added advantage of providing a large series of cases with the same affection.

The venous drainage of the brain in children with spina bifida aperta showed great variability. The anomalous findings are described and discussed (Chapters IV and V).

The chief findings are:

1) The skull in spina bifida aperta shows the normal number and configurations of the jugular foramen and emissaries. The drainage via the vertebral column seems to be enhanced.

2) At the side of the anterior fontanelle there were transverse collaterals in the falx having connections with the anterior and posterior cerebral veins and anastomosing with the deep cerebral veins by the posterior cerebral vein.

There were also clear abnormalities of the falx and the inferior longitudinal sinus. The great vein of Galen/straight sinus showed two courses. The great vein of Galen was several times longer. The

straight sinus was short and took a more vertical direction.
3) In several specimens the ascending cerebral veins changed direction close to the sinus, but sometimes only in the wall of the longitudinal sinus. In some cases this change of direction was so extreme that instead of emptying into the sinus in the frontal direction the vein turned sharply to enter the sinus in the direction of the torcular, or *vice versa*. The course of the ascending cerebral veins were directed frontally on the right side in 63 per cent of the cases and on the left side in 58 per cent of the cases. The number of the stems of the ascending cerebral veins was not increased.
4) The lateral sinuses ran near to the foramen magnum to the sigmoid sinuses. At the origin of the sigmoid sinus there was a confluence receiving the petrosal veins and veins and sinuses from the floor of the middle fossa and from the cerebral hemispheres. The embryonic tentorial sinus usually persists in these children. The inferior vermian vein draining the Arnold-Chiari malformation had close connections with the margin of the foramen magnum. During elevated intracranial pressure the inferior vermian vein is pressed against the dorsal margin of the foramen magnum, thus producing haemorrhagic infarction and eventual subsequent necrosis of the cerebellar tonsils. The petrosal vein drained more laterally. The dura of the posterior fossa consisted of a plexiform structure in which no clear distinction could be made between the various sinus systems normally present in the dural layers of the posterior fossa, thus constituting an occipital plexus. The walls of the sinuses were weak and were vulnerable to mechanical pressure.
5) The foramen magnum of the skulls of children with spina bifida aperta was larger than in normal children. The distance of the torcular to the ventral margin of the foramen magnum was smaller in spina bifida aperta. This distance remained unchanged at least until 9 months of age in normal children as well as in children with spina bifida aperta, although there was an increase in size of the foramen magnum in both with advancing age.
The type of the abnormality of the cerebral venous system did not, however, appear to be related to the degree of hydrocephalus.

In children with spina bifida aperta there is an increase in the drainage of blood from the brain to the external jugular system, and, since this is relieved by reducing intracranial pressure, it would

appear to be a secondary phenomenon.

The improved collateral circulation explains not only the dilatation of the superficial cranial veins in the presence of elevated intracranial pressure but also the width of the common facial vein.

If the venous anomalies actually contribute to the development of hydrocephalus in cases of spina bifida aperta, haemodynamic factors are probably responsible. Although the pathological findings reported here concern only the anatomical situation and no pressure measurements are included, certain conclusions can nevertheless be drawn. The nature of the abnormalities found in the venous structures makes it likely that increased CSF pressure, which in these children is more likely to obstruct the venous outflow than in normal children, can cause acutely an elevation of the intracranial venous pressure.

There is evidence that increased CSF pressure results in increased cerebral venous pressure and that increased cerebral venous pressure can also produce an increased CSF pressure, so that these two can constitute a vicious circle, interrupted when a ventriculocardiac shunt is applied. Thus, it is important to maintain a constant, normal intracranial venous pressure. At present this can only be produced by a well-functioning shunt that drains the CSF extracranially.

No surgical procedure on the cranial venous system to alleviate the parallel dependences of the cranial venous pressure on the CSF pressure in children with spina bifida aperta is yet available.

More must be known about the exact level of pressure in different parts of the venous system in the heads of these children before a logical approach can be made to tackling the vicious circle of the interaction between the CSF pressure and the intracranial venous pressure.

SAMENVATTING

Het intracraniële veneuze systeem werd bij kinderen met een spina bifida aperta macroscopisch onderzocht. De aandacht hierop werd gevestigd doordat de oppervlakkige venen van de schedel zo gezwollen waren, indien progressieve hydrocephalus bestond. Bovendien werd tijdens het aanbrengen van een ventriculo-cardiale drainage voor de behandeling van de hydrocephalus vaak een opvallend dikke vena facialis communis gezien.

Bij de aanvang van deze studie werden preparaten van hydrocephale kinderen onderzocht, zonder dat wij ons bekommerden om de oorzaak van de hydrocephalus. Toen bij kinderen met een spina bifida aperta regelmatig ernstige afwijkingen werden gevonden, beperkten wij ons onderzoek tot deze gevallen. Bovendien bood deze beperking als voordeel, dat wij een grote groep konden onderscheiden, die een gelijke aandoening had.

De veneuze drainage van het cerebrum van kinderen met een spina bifida aperta toonde een grote variatie ten opzichte van de normale anatomie. De abnormale bevindingen werden beschreven in hoofdstuk IV en besproken in hoofdstuk V. De vorm van de afwijking leek echter geen verband te houden met het wel of niet optreden van hydrocephalus.

Indien de veneuze anomaliën al enig aandeel in het optreden van hydrocephalus bij spina bifida aperta hebben, zal dit eerder door hemodynamische factoren bepaald worden. In deze studie werden geen drukmetingen vermeld, doch slechts pathologisch anatomisch bevindingen. Toch zijn daaruit wel bepaalde konklusies te trekken. De abnormale bouw van de veneuze structuren maakt wel waarschijnlijk dat verhoogde intracraniële veneuze druk veroorzaakt kan wor-

den door verhoogde liquordruk in het ventrikelsysteem bij deze kinderen. Dit heeft op zijn beurt invloed op het in stand houden van de verhoogde liquordruk in het ventrikelsysteem. Verder is het opvangen van de arteriële polsgolf door het verminderen van de windketelfunctie niet in gelijke mate aanwezig als bij gezonde kinderen. Dit zal medebepalend zijn voor de beschadiging van de hersenstructuren.

De verbetering van de collaterale circulatie verklaart waarom tijdens verhoogde intracraniële druk de schedelhuidvenen zo gevuld zijn. Ook een dikke vena facialis communis is om gelijke redenen begrijpelijk. Men kan dan ook stellen dat bij kinderen een spina bifida aperta het jugularis externa systeem van groot belang is voor de drainage van het bloed van het cerebrum. Bij kinderen met een hydrocephalus is er een toename van de drainage van bloed uit de hersenen via het jugularis externa systeem. Aangezien dit opgeheven wordt doordat de intracraniële druk wordt verlaagd, is deze toename van de drainage via het jugularis externa systeem een secundair effect.

Er zijn aanwijzingen dat verhoogde liquordruk een verhoogde veneuze druk van het cerebrum veroorzaakt. Bovendien beinvloedt de cerebrale veneuze druk eveneens de liquordruk, zodat deze twee een circulus vitiosus vormen, die doorbroken wordt, doordat een ventriculo-cardinale shunt wordt aangebracht. Derhalve is het belangrijk om bij deze kinderen een constante en normale intracraniële veneuze druk te handhaven. Op het ogenblik is dit alleen mogelijk doordat de liquordruk normaal wordt gehouden door middel van een operatie waarbij de liquor naar buiten het hoofd wordt afgeleid. Er is nog geen operatie aan het veneuze systeem bekend die de abnormale afhankelijkheid van de veneuze druk van de liquordruk vermindert. Meer kennis is vereist van de drukverhoudingen in het veneuze systeem in het hoofd van deze kinderen voordat op logische wijze de circulus vitiosus van de interactie tussen liquordruk en intracraniële veneuze druk operatief doorbroken kan worden.

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