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Hydrocephalus in craniosynostosis: a review

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Abstract *Introduction:* Ventricular dilatation in the presence of primary craniosynostosis is a unique condition with respect to pathogenesis, clinical significance, and morphological appearance. It is rarely observed in nonsyndromic craniosynostosis, and in these cases usually attributable to coincidental disorders. Conversely, it is a common feature of syndromic craniosynostosis, affecting at least 40% of patients with Crouzon's, Pfeiffer's or the Apert syndrome. Shunt-dependent hydrocephalus is predominantly associated with Crouzon or Pfeiffer syndrome while in the Apert syndrome the usual finding is nonprogressive ventriculomegaly which, however, may also occur in some cases of Crouzon syndrome. *Pathogenesis:* The pathogenesis of progressive hydrocephalus remains somewhat obscure, a hypoplastic posterior fossa and a venous outlet occlusion at the skull base being the main causative factors discussed in literature. Ventriculomegaly may reflect primary brain maldevelopment or in some cases even a compensated state of increased cerebrospinal fluid (CSF) outflow resistance. *Clinical evaluation:* Clinical evaluation is mainly aimed at identifying progressive hydrocephalus, but diagnosis is hampered by the fact that classical clinical signs may be absent, and that ventricular dilatation will often become evident only after decompressive cranial surgery. Moreover, mild ventriculomegaly may in some cases coexist with intracranial hypertension from craniostenosis. Therefore, careful monitoring of intracranial pressure and ventricular size in the pre- and postoperative period is a diagnostic mainstay. *Conclusion:* In true hydrocephalus ventriculo-peritoneal shunting is currently the single promising mode of treatment.

Keywords Cranioynostosis · Crouzon's syndrome · Hydrocephalus · Venticulomegaly · Chiari malformation · Venous outflow obstruction

Introduction

Hydrocephalus accompanying craniosynostosis is a remarkable condition with regard to pathogenesis, clinical significance, clinical and radiological appearance, and treatment. It has been appreciated since the early days of craniofacial surgery [49, 53], and has been recognised as a disorder to be clearly distinguished from craniosynostosis secondary to chronically shunted hydrocephalus [33]. For decades, owing to the limitations of imaging techniques, knowledge about the condition was mainly based on incidental cases [2, 15, 30, 31, 37, 40] or on small, highly selected series [18]. Some of the classical clinical studies on craniosynostosis did not mention hydrocephalus at all [60]. Only in the 1980s did the widespread use of computerised tomography and magnetic resonance imaging

permit a more systematic study, addressing the questions of incidence, pathogenesis, and clinical significance of dilated ventricles in craniosynostosis [4, 13, 20, 48]. From then on, numerous additional reports documented an increasing interest in this issue [7, 8, 10, 41, 46, 51, 52, 55].

Up to now, there has been agreement in little more than that:

- Enlarged ventricles in craniosynostosis may represent either shunt-dependent hydrocephalus or shunt-independent ventriculomegaly [8, 10, 13, 48]
- Progressive hydrocephalus is mainly a problem of complex craniosynostosis [8, 13, 20, 48],
- Hydrocephalus in primary craniosynostosis poses a particular diagnostic problem, since both disorders may elevate intracranial pressure, while obviously exerting opposite forces on the intracranial space: expansion versus constriction [8, 13, 20]

Consequently, the main issues to be addressed in this review will be:

- The causative mechanisms of both progressive hydrocephalus and nonprogressive ventriculomegaly
- The criteria differentiating between the two conditions
- The options for treatment and their interference with surgery for synostosis

Epidemiology

The reported figures of both progressive hydrocephalus and nonprogressive ventriculomegaly in craniosynostosis vary to a great extent, and we can only speculate about the reasons, which may include different referral patterns, different nosologic classification, and different diagnostic criteria of progressive hydrocephalus, since separation from nonprogressive ventriculomegaly is a less than simple matter [8, 13, 18]. In addition, inaccurate definition of what is referred to as "ventricular dilatation" in contrast to normal conditions, and even sometimes confusion of mild ventricular enlargement with simple ventricular distortion in a deformed, towering skull may contribute to this variety [23]. Finally, regarding the dynamic nature of hydrocephalus, accurate figures on its prevalence may only be derived from prospective longitudinal studies, including repeated imaging prior to and after craniosynostosis surgery. While a single prospective study on a small number of patients has indeed been reported [48], no comprehensive longitudinal investigation has, as yet, come to the authors' attention.

Irrespective of these difficulties, in many studies a striking difference between simple monosutural craniosynostosis and the complex craniofacial syndromes has been consistently reported [8, 13, 20, 48, 66]. In the former any ventricular enlargement is an unusual finding, and shunt-dependent hydrocephalus is even less often encountered, its incidence being comparable to that of the general population unaffected by craniosynostosis (Table 1) [8]. Conversely, ventricular dilatation of either origin has been reported in 30 to 70% of patients with Crouzon's and Pfeiffer's syndrome [8, 40, 48, 51], and in 40 to 90% of Apert patients [11, 25, 52] (Table 2). Also, it has been consistently reported that shunting proved necessary predominantly in Crouzon's and Pfeiffer's syndrome, while in the Apert syndrome most cases of enlarged ventricles remained stable without a shunt [25, 48, 52]. Patients with Saethre–Chotzen syndrome or Muenke syndrome appear to be rarely affected by dilated ventricles, and shunt-dependent hydrocephalus has so far not been documented. As the diagnosis of these latter syndromes has only recently been improved by implementing molecular techniques, earlier studies should be looked at with due scepticism. We, for

Table 1 Ventricular size and prevalence of true hydrocephalus in patients with various types of craniosynostosis examined at the authors' institution. *NA* not applicable

Type of craniosynostosis	Sample size	Ventricular dilatation (patients)			Shunt-dependent (patients[a])	Age at shunting minimum, median, maximum (months)
		Mild	Moderate	Severe		
Sagittal	133	8	3	1	2 (a)	1, N.A., 8
Frontal	88	8	1	2	1 (b)	2
Unicoronal	54	0	1	2	2 (c)	0.1, N.A., 0.2
Bicoronal	25	1	1	2	3 (d)	0.1, 0.3, 2
Multiple sutures	15	2	0	0	0	N.A.
Total isolated	*315*	*19*	*6*	*7*	*8*	N.A.
Crouzon's[b]	63	15	11	6	10	2, 20, 374
Pfeiffer's	15	2	4	6	9	3, 6, 78
Apert	45	25	7	0	2	18, N.A., 42
Saethre–Chotzen	37	3	0	0	0	N.A.
Muenke	24	2	0	0	0	N.A.
Total syndromic	*184*	*47*	*23*	*11*	*20*	N.A.

[a]Aetiology of shunted hydrocephalus: *a* posthaemorhagic (1 patient), tonsillar herniation (1), *b* cerebral malformation (1), *c* myelomeningocele (2), *d* myelomeningocele (2), amniotic band sequence (1)

[b]Two cases of FGFR3-related Crouzon's syndrome (with acanthosis nigricans) included

Table 2 Reported prevalence of nonprogressive ventriculomegaly and progressive hydrocephalus in the common types of complex craniosynostosis syndrome

Reference	Patient (*n*)	Normal (%)	Ventriculomegaly (%)	Hydrocephalus (%)
Crouzon's				
[48]	12	50	33	17
[51]	35	29	63	9
[8]	86	58	16	26
Present study	63	49	35	16
Pfeiffer's				
[48]	5	60	0	40
[41]	11	9	27	64
[8]	18	72	0	28
Present study	15	20	20	60
Apert				
[11]	28	0	93	7
[46]	25	40	48	12
[25]	13	0	92	8
[52]	60	57	35	8
Present study	45	24	72	4

instance, erroneously classified a case of nonprogressive ventriculomegaly as Saethre–Chotzen syndrome in our own study [13]. Hydrocephalus may also occur in some rare craniosynostosis syndromes, including the FGFR3-associated Crouzon's syndrome with acanthosis nigricans [45], one of two of our own patients needed a shunt, as well as Carpenter syndrome [48, 62], Antley–Bixler syndrome [34], Shprintzen–Goldberg syndrome [22], and some other rare syndromes [1, 3, 68] (Table 3). In addition, hydrocephalus is usually present in thanatophoric dysplasia in which craniosynostosis is part of the typical phenotype [12]. Progressive hydrocephalus appears to be related to the number of fused sutures and to the time of fusion [7, 9]. It does not appear to be related to a specific skull shape except the Kleeblattschädel deformity, which in most cases is associated with progressive hydrocephalus [8, 20, 59]. Recently, a remarkable concurrence of coronal synostosis with hydrocephalus related to neural tube defects has been reported [39]. Four own cases of myelomeningocele affected with either unilateral (2 patients) or bilateral coronal synostosis (2 patients) seem to corroborate this observation. However, a large comprehensive French study did not mention this particular association at all [8].

Aetiology and pathogenesis

In craniosynostosis progressive hydrocephalus is conceivable as the result of either CSF outflow obstruction or malabsorption, both being directly or indirectly related to the osseous pathology or to coincidental disorders independent from craniosynostosis. Hydrocephalus may be rapidly or slowly progressive due to a more or less elevated intracranial pressure or it may even become compensated leaving the intracranial pressure at a normal level. Impaired CSF absorption due to venous sinus hypertension typically causes general enlargement of the inner and outer CSF spaces if the skull is still capable of expanding, or it induces a pseudotumour-like state in a non-expanding skull [32, 50, 61]. Conversely, nonprogressive ventriculomegaly may be attributed to primary brain maldevelopment or to secondary involution of brain parenchyma, i.e. brain atrophy. All these conditions have been suggested to play a role in craniosynostosis.

In isolated monosutural craniosynostosis the few cases of progressive hydrocephalus can almost always be attributed to coincidental disorders independent from craniosynostosis, which included ventricular haemorrhage, meningitis, aqueductal stenosis, and neural tube defects [8, 13, 18, 20, 48]. Yet a few cases of isolated craniosynostosis present with an intracranial pathology usually known from complex craniosynostosis, e.g. tonsillar herniation (Table 1) [8]. In these cases the lambdoid suture seems to be most commonly involved, either separately or combined with sagittal suture synostosis [44, 54]. Nonprogressive ventriculomegaly is also most often attributable to coincidental disorders like perinatal haemorrhage resulting in a compensated hydrocephalus, or to dysgenetic cerebral anomalies, which may account for most cases of ventriculomegaly in trigonocephaly [14].

The causative mechanism of progressive hydrocephalus in syndromic craniosynostosis is the subject of continuing discussion, and reported data are in part conflicting. The idea of coincidental disorders has now largely been abandoned, although aqueductal atresia has been reported in

Table 3 Types of craniosynostosis in which nonprogressive ventriculomegaly and progressive hydrocephalus are considered common features

Craniosynostosis type	Reference(s)
Pfeiffer's syndrome	[8, 20, 41]
Crouzon's syndrome	[8, 51]
Crouzon's syndrome with acanthosis nigricans	[45]
Apert syndrome	[11, 25, 52]
Beare–Stevenson cutis gyrata syndrome	[68]
Thanatophoric dysplasia	[12]
Carpenter syndrome	[48, 62]
Shprintzen–Goldberg syndrome	[22]
Antley–Bixler syndrome	[34]
Jones craniosynostosis—Dandy–Walker syndrome	[3]
Kleeblattschädel deformity	[59]
Amniotic band sequence	[1]

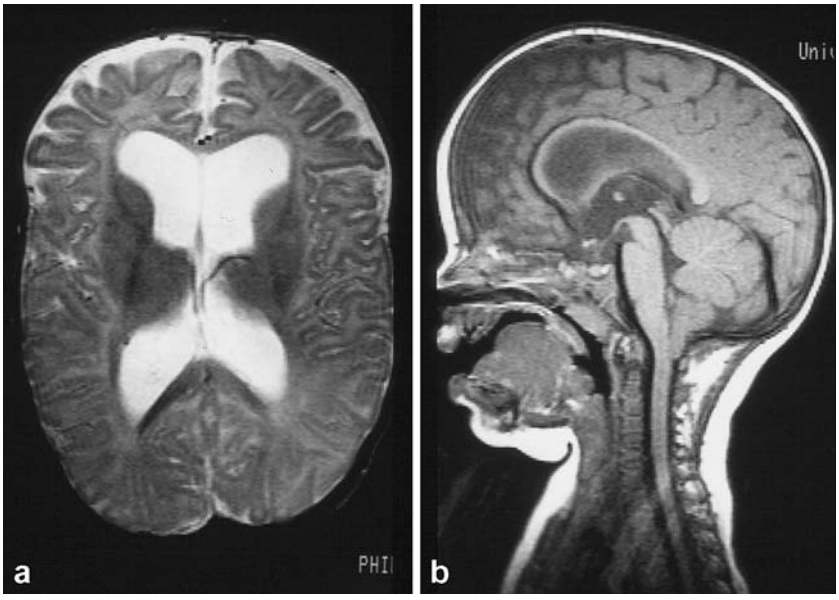

Fig. 1 **a**, **b** MRI of a 6-month-old girl with Crouzon's syndrome, showing herniated cerebellar tonsils and dilated ventricles that had been normal on ultrasound at the age of 2 months. Primary shunting was carried out because of rapidly progressive ventricular dilatation and papilloedema

exceptional cases [18]. At present, most authors believe in one of two pathogenic factors or a combination of both:

- Mechanically increased CSF outflow resistance due to constriction of the posterior fossa [7, 9, 13, 15, 19, 64]
- Impaired CSF absorption resulting from venous outflow obstruction [8, 19, 55]

The first theory was proposed by Rieping [53] as early as 1919, and was later pursued by David and coworkers [15], and by Venes [67]. In fact, with few exceptions most cases of hydrostatic hydrocephalus exhibit compromised CSF spaces of the posterior fossa, a small fourth ventricle, and a Chiari-like anomaly (Figs. 1, 2) [7–9, 19, 64]. Correspondingly, tonsillar herniation of varying degrees is commonly observed in Crouzon's and Pfeiffer's syndromes, but rarely found in the Apert syndrome [7, 66]. In our own series an early and rapidly progressing hydrocephalus was frequently though not invariably associated with a more severe ectopia of the cerebellar tonsils. The anomaly was absent in only three patients who all presented with slowly progressive hydrocephalus. Crowding of the posterior fossa appears to be an acquired disorder secondary to deficient occipital cranial expansion and results in a condition that has been referred to as "cephalocranial disproportion" [28]. It has been related to the time of fusion of the lambdoid suture, which in Crouzon's and Pfeiffer's syndromes is completed at an earlier age than in the Apert syndrome [7]. Support for this view comes from

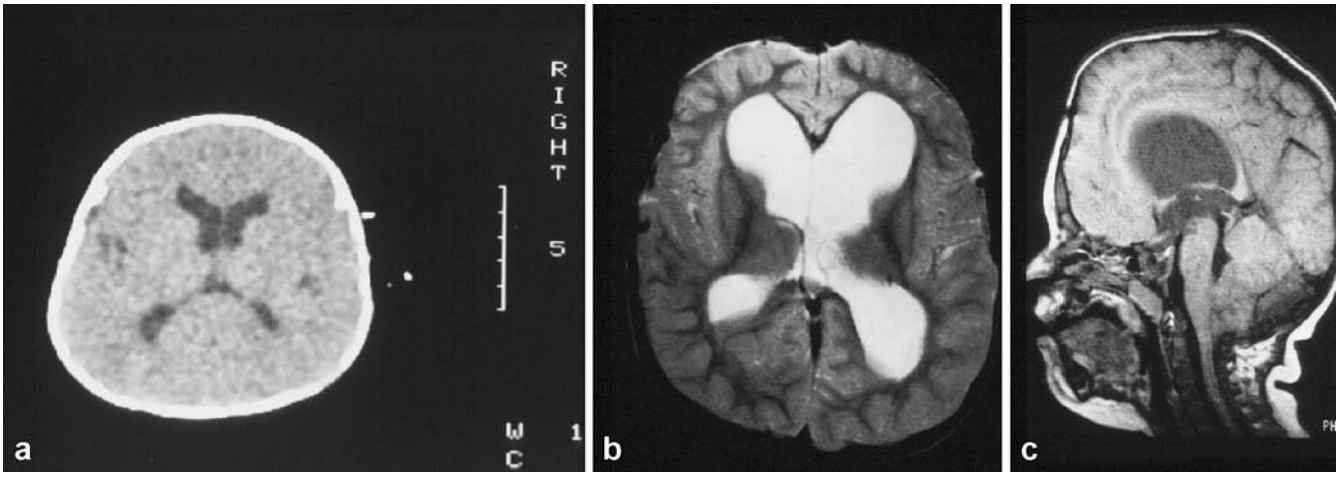

Fig. 2 **a** Preoperative CT of a boy with Pfeiffer's syndrome prior to forehead advancement at the age of 6 months. **b** MRI at the age of 30 months showed marked ventricular dilatation in the presence of papilloedema, which prompted ventriculo-peritoneal shunting. **c** Note crowding of the posterior fossa and the patent aqueduct

experimental findings [38], from estimates of the posterior fossa volume [58, 64], and from well-documented cases of postnatally developing tonsillar herniation [6, 29, 54, 65]. CSF pathways are considered to be compromised at the site of the extracerebral cisterns [9, 19, 64], and findings derived from isotope cisternography seem to confirm this view [57]. Others believed in an underlying aqueduct stenosis, probably because of the CT appearance of the compromised fourth ventricle on CT [48, 51], but MR imaging usually shows the aqueduct open (Figs. 1, 2) [7, 8, 64]. The concept of mechanical CSF outflow obstruction may be called into question since in a few cases of progressive hydrocephalus hindbrain herniation is missed [8], while it is present in many other cases not affected by hydrocephalus (Fig. 3) [8, 63]. The theory is also challenged by the fact that posterior fossa decompression failed to sufficiently restore normal CSF circulation [9]. Therefore, constriction of the posterior fossa may not be the single mechanism causing hydrostatic hydrocephalus in craniosynostosis.

The hypothesis of defective CSF resorption due to impaired venous outflow was initially proposed by Hoffman and Hendrick [27], but essentially the same mechanism had been postulated decades before [23, 24]. It was the craniofacial group of the Necker Hospital of Paris who advanced the concept of venous sinus hypertension as a major factor involved in progressive hydrocephalus [8, 55]. Similar to the conditions found in achondroplasia [50, 61] Sainte-Rose and colleagues documented in some hydrocephalic Crouzon's patients a fixed venous sinus hypertension caused by a stenosis of the jugular foramen [55]. Moreover, in one of these patients they demonstrated normalisation of CSF pressure and ventricular size after performing a venous bypass between the transverse sinus and the jugular vein. Later on, in a comprehensive study, Cinalli and coworkers [8] angiographically demonstrated jugular foramen stenosis as well as an extensive venous collateral network in many Crouzon's patients and in a few Apert patients. Most of them actually had progressive hydrocephalus while only few hydrocephalic patients presented with a normal venous outflow. By applying the vascular theory the authors provided arguments why progressive hydrocephalus is usually limited to early childhood in that they assumed that venous collateral channels will progressively open with increasing age, and, hence, permit gradual normalisation of the venous sinus pressure. In addition, their finding that the ventricles dilated only in cases in which at least some sutures were still open, complies with the concept of venous hypertension as in a totally rigid skull the latter should rather induce a pseudotumour-like state [32]. Meanwhile, venous hypertension has been accepted as a major pathogenic mechanism by other investigators too [19, 54, 64], although some questions still remain. Given that the same mechanism (CSF malabsorption) accounts for hydrocephalus in Crouzon's syndrome and in achondroplasia, why do patients with achondroplasia only rarely need a shunt, and why do patients with Crouzon's syndrome only rarely present with enlarged subarachnoid spaces, which are so common in achondroplasia? In addition, many patients presenting with severely elevated intracranial pressure and an extensive collateral venous network, actually have normal ventricles [63]. Finally, although the successful treatment of hydrocephalus by means of a venous bypass provided a strong argument in favour of an underlying venous problem it could unfortunately not be proven with further patients, since technical problems and the time needed for the small vascular graft to become an efficient shunt prevented further attempts at this kind of treatment [8].

In view of the fact that most patients with progressive hydrocephalus simultaneously exhibit signs of both venous outflow obstruction and crowded posterior fossa, most authors currently favour a combined action of both mechanisms by assuming that venous hypertension causes a CSF absorption deficit as well as brain swelling resulting in

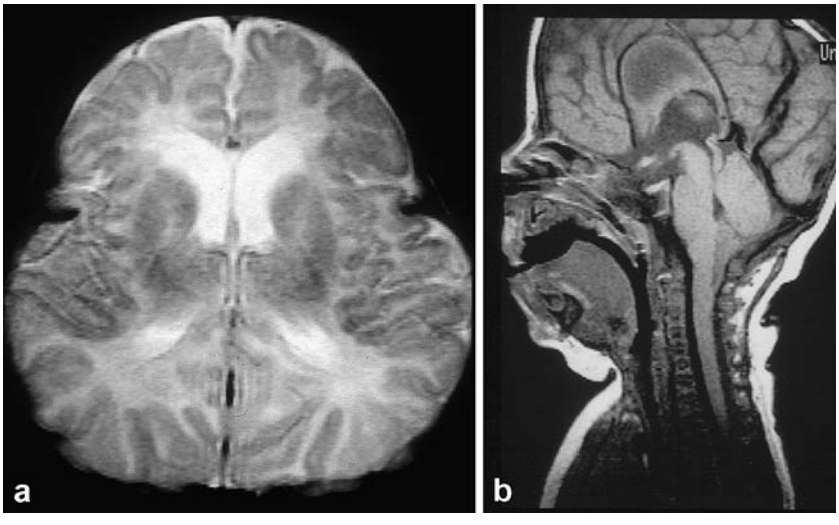

Fig. 3 **a**, **b** Magnetic resonance image of a 3-month-old girl with Pfeiffer's syndrome showing mild ventricular dilatation, which remained stable after cranial reconstruction despite severe hindbrain herniation. No shunting was required during the follow-up of 21 months

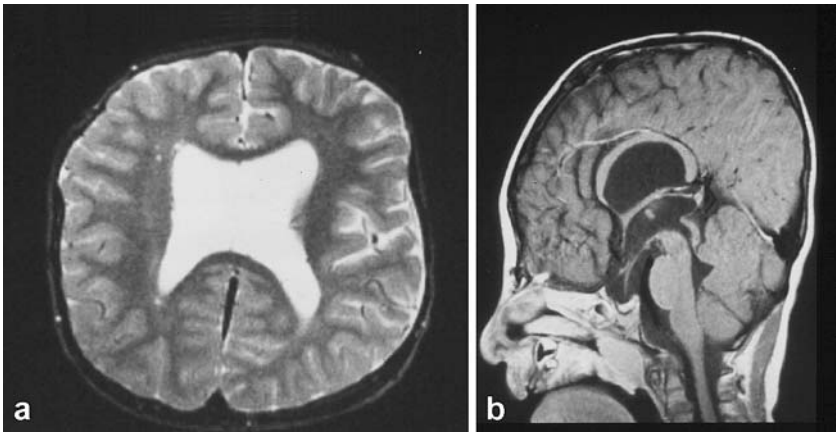

Fig. 4 **a**, **b** Magnetic resonance image of a 4-year-old boy with the Apert syndrome prior to a repeated cranial expansion because of headaches and bulging craniectomy sites. Postoperatively, the ventricles remained essentially unchanged, and no shunting was required during the following 10 years

tonsillar herniation [19], or that it aggravates the pre-existent cephalo-cranial disproportion by venous engorgement [8, 64]. The latter appears to be the most accepted theory at present.

While there is little doubt that nonprogressive, shunt-independent ventriculomegaly really exists in a number of cases with complex craniosynostosis, opinions as to the underlying pathology differ. Brain atrophy has been postulated by some authors [2], and on the basis of our own observations it does appear to play a role in a few cases [13]. The striking frequency of mild to moderate nonprogressive ventricular dilatation in the Apert syndrome, which so often concurs with other cerebral abnormalities clearly suggests primary brain maldevelopment (Fig. 4) [10, 48, 66]. Cohen and Kreiborg used the term "distortion ventriculomegaly" to emphasise the malformative nature of this kind of ventricular enlargement [10]. However, shunt-independent ventriculomegaly is also observed in Crouzon's syndrome, which is usually associated with normal cerebral development (Fig. 5). Therefore, Cinalli and coworkers [8] favour the idea of a compensated hydrocephalic state, which they attribute to the same, though less

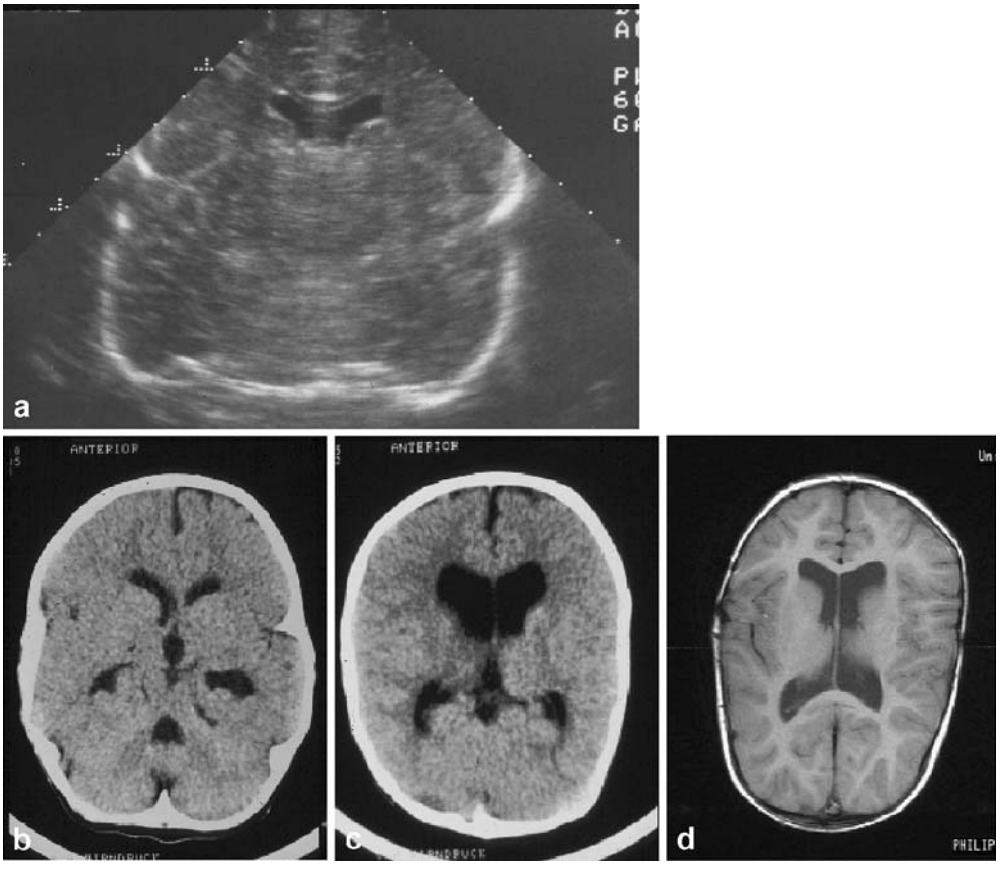

Fig. 5 **a** Sonographic picture at 4 weeks of age, **b**, **c** CT at 14 months, and **d** MRI at 44 months of a girl with Crouzon's syndrome who received two cranial reconstructive procedures at the ages of 6 weeks and 16 months. In both instances intracranial hypertension (tense fontanel and papilloedema respectively) was successfully treated without shunting. The follow-up was 12 years

active, mechanisms responsible for progressive hydrocephalus. As there is little doubt that compensated hydrocephalus really exists in some cases it may be concluded that different mechanisms may contribute to nonprogressive ventriculomegaly.

Finally, there are reports on pericerebral CSF collections developing after cranial surgery, which in some cases prompted treatment with a shunt [8, 46]. This implied the idea of CSF malabsorption, probably due to the same mechanism as in progressive hydrocephalus, i.e. venous hypertension [8, 46]. Several investigations including intrathecal infusion tests [17, 36] and transcranial Doppler sonography [47] seemed to support this idea, although these studies predominantly addressed intracranial pressure and compliance rather than selectively assessing CSF absorption. Consequently, other authors have questioned attributing prominent subarachnoid spaces to a defective CSF absorption [5]. As a matter of fact, local enlargement of subarachnoid spaces is a quite common feature of craniosynostosis even before surgical intervention (Figs. 6, 7) [5, 26]. It is mainly confined to regions of compensatory skull expansion, and is found to be most pronounced in scaphocephaly [26]. The possible underlying mechanisms have been discussed by several investigators [5, 8, 26, 56]. In view of modern concepts of calvarial growth in craniosynostosis it is conceivable that subarachnoid spaces passively dilate to accommodate for local compensatory skull expansion caused by an intrinsic disparate bone growth [16, 42, 43]. Chadduck et al. [5] suggested that the fluid collections may then exert an additional expanding force to the bone by "an augmented transmission of the brain pulsations". In addition to this theory some relationship to the so-called "benign enlargement of subarachnoid spaces" or "external hydrocephalus" has been discussed in view of the fact that prominent subarachnoid spaces are usually confined to patients less than 1 year of age [5, 56]. In any case, pericerebral fluid collections of major clinical significance, i.e. necessitating specific treatment, will obviously remain an exceptional finding [8], and was not once observed at our own institution, while transient enlargement of the subarachnoid space without elevated pressure is a common and well-known phenomenon after expanding skull surgery at any age [8, 13, 37, 42, 43].

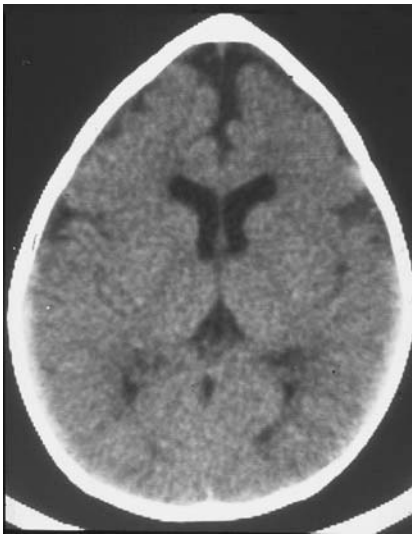

Fig. 6 Computerised tomography scan of a 7-month-old boy with isolated trigonocephaly showing local enlargement of the frontal subarachnoid space. There were no signs of intracranial hypertension

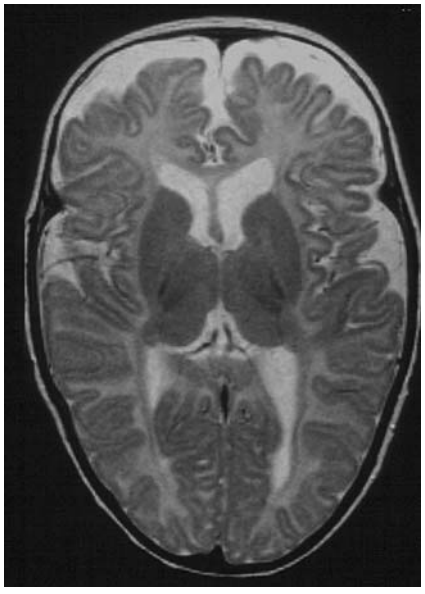

Fig. 7 Magnetic resonance images of a 5-month-old girl with isolated sagittal synostosis showing extracerebral fluid collections over the frontal lobes. There were no signs of intracranial hypertension

In summary, patients with complex craniosynostosis, in particular Crouzon's, Pfeiffer's, and the Apert syndrome, may present with different kinds of enlarged CSF spaces—progressive hydrocephalus, nonprogressive ventriculomegaly and dilated subarachnoid spaces. Clinical evaluation will have to focus on selecting those cases that require shunt treatment.

Diagnostic evaluation and indication for treatment

Shunt-dependent hydrocephalus in childhood is usually recognised on the basis of progressive ventricular enlargement and signs of elevated intracranial pressure. In craniosynostosis the diagnosis is less easily established due to the fact that in a considerable proportion of shunt-dependent patients the hydrocephalic condition appears obscured within the rigid synostotic skull only to become visible as ventricular dilatation after decompressive cranial surgery [8, 13]. Conversely, intracranial hypertension is an ambig-

uous sign that may reflect increased CSF outflow resistance or venous hypertension or the constricting forces of the osseous pathology [64]. Even mild or moderate ventricular enlargement in the presence of intracranial hypertension does not necessarily signify shunt-dependent hydrocephalus (Figs. 3, 4, 5). In contrast to other authors [8, 48] we do not recommend relying on accelerated head growth or radiological signs of hydrostatic hydrocephalus such as dilated temporal horns or periventricular lucency as the significance of these signs turned out to be low in our patients (Fig. 5). Apart from the fact that marked ventricular dilatation usually indicated shunt dependence in our series, ventricular size and shape did not provide reliable criteria for the differentiation between nonprogressive ventriculomegaly and true hydrocephalus.

In spite of these diagnostic problems some guidelines for use in clinical practice can be established. To begin with, ventricular distortion in a towering skull should clearly be separated from true dilatation [23]. Thereafter, the individual risk of developing hydrostatic hydrocephalus may be estimated by looking for the main factors signalling a high risk: complex syndromic craniosynostosis, in particular the Crouzon and Pfeiffer phenotypes, Kleeblattschädel deformity, progressive synostosis involving the lambdoid suture at an early age, and a crowded posterior fossa with tonsillar herniation. Routine MR imaging including MR venography is recommended in all patients suffering from Crouzon's, Pfeiffer's or the Apert syndrome [8, 63, 66]. In case of tonsillar herniation they should be kept under surveillance with regard to both intracranial pressure and ventricular size, making use of ophthalmoscopy and possibly ultrasound as basic diagnostic tools. As in other forms of hydrocephalus subjective symptoms of intracranial hypertension like headaches, nausea and vomiting may be missed, while, for instance, papilloedema was a rather common sign in our patients.

In some 50% (in our study group 33%) of the patients affected by hydrostatic hydrocephalus the diagnosis is straight-forward because of rapidly progressive ventricular dilatation prior to any surgical intervention, and this is to be expected mainly during infancy (Fig. 1) [8]. In most of the remaining patients ventricular dilatation of varying degrees will only develop following decompressive surgery for craniosynostosis. In these cases the indication for shunting is mainly based on severe ventricular dilatation or evidence of persisting intracranial hypertension, which is ascertained by papilloedema or by direct pressure monitoring (Fig. 2). However, it is important to remember that after cranial surgery the artificially created space is quite often accommodated by some enlargement of the intra- and extracerebral CSF spaces, and that papilloedema may need several weeks to subside. In addition, the possibility of a compensated or even slowly progressive hydrocephalic state should also be taken into account. Therefore, it is prudent to keep these patients under long-term surveillance.

There are a few patients left who present with mild or even moderate, sometimes slightly progressive ventricular enlargement and concurrent signs of elevated intracranial pressure (Figs. 3, 4, 5). Although these patients meet the classical criteria of hydrostatic hydrocephalus, some of them actually will remain shunt-independent after adequate cranial expansion [13, 21]. Therefore, in the absence of reliable diagnostic criteria it may be appropriate to operate on the craniosynostosis first and to carefully observe the development of ventricular size and intracranial pressure afterwards. Long-term surveillance is advised in these cases as well [8].

Treatment and outcome

Although improvement of ventricular dilatation has been anecdotally reported following removal of constricting bony ridges [59] most attempts to relieve progressive hydrocephalus by eliminating its putative causes have failed or could not be established as routine techniques [7, 8]. To the authors' knowledge, third ventriculostomy has not been evaluated within this context, and a single, successful procedure in a personal case of progressive hydrocephalus associated with tonsillar herniation is not conclusive. Hence, shunting appears to be the single feasible mode of treatment. In the presence of hindbrain herniation there is certainly no place for the lumbar route as this can lead to a fatal outcome [6]. When selecting the valve system it seems advisable to take particular precautions against overdrainage, since the latter may induce a pseudotumour-like state of venous origin [32], which in craniosynostosis may add to the pre-existing venous problem.

If it is accepted that venous outflow obstruction contributes to the pathogenesis of hydrocephalus it is an intriguing concern that shunting does not address the vascular pathology. In fact, according to several investigators, impaired venous outflow may account for continuing intracranial hypertension at least throughout early childhood until sufficient venous collaterals have opened at the age of about 6 years [19, 21, 54, 65]. However, the clinical significance of untreated venous outflow obstruction is unclear, and, to the authors' knowledge, the particular impact of a functioning shunt on venous hypertension within a synostotic skull has not been investigated in detail.

It may be tempting to treat any kind of ventricular dilatation by shunting with the simple intention of gaining space and avoiding extensive cranial surgery. Without regarding the problem of shunt morbidity this policy may be justified at best occasionally after brain growth has been completed. This was the case in an adult Crouzon's patient with moderate ventricular dilatation whose chronic head-

aches improved after receiving a shunt at another institution. But it is certainly ill-advised in childhood, since the drained CSF spaces will rapidly be filled up by the growing brain while at the same time the skull is deprived of an important growth stimulus.

The timing of shunting in relation to surgery for synostosis is a matter of additional concern since the stability of cranial reconstruction may be endangered if the dural envelope does not rapidly expand because of the artificial depletion of CSF spaces, and the cranial content fails to support the bone plates in due time. One of our patients was reoperated because skin tension caused a displacement of some bone segments that had not been sufficiently secured during the initial procedure. In view of this risk, some authors recommended giving shunting priority over cranial remodelling [37], which, however, will mean unnecessary shunt operations in certain cases. Therefore, taking the dynamics of bone healing and brain growth into account, it is reasonable to postpone any reconstructive cranial procedure after primary shunt insertion until intracranial hypertension recurs. Secondary shunting after cranial remodelling should be deferred for at least 2 months. If this is not feasible, every effort should be made to secure the surgical bone plates prior to impending shunt surgery.

There is little evidence that dilated ventricles per se have an adverse effect on mental outcome [46, 48, 52] except that the severe congenital hydrocephalus, as observed in the Kleeblattschädel deformity obviously carries an increased risk of a subnormal performance level [35, 59]. As in other hydrocephalic states the prognosis mainly depends on coincidental cerebral abnormalities and on the detrimental effect of long-standing elevated CSF pressure. Dealing with this latter problem is the cardinal objective of treatment.

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