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Surgery for Chiari 1 malformation: the Lille experience

Matthieu Vinchon [1,2]

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Abstract
Purpose Decision-making in chronic tonsillar herniation (CTH) in children is complicated because many cases are diagnosed incidentally; on the other hand, its clinical impact may be underestimated. Furthermore, its surgical management is controversial.
Methods In the present review, we tried to design a semi-quantitative approach to diagnosis, defining presenting symptoms as compatible, suggestive, or differential diagnoses. We expose our rationale for surgery. We review our experience with extensive posterior fossa decompression (PFD) with systematic dural opening and low threshold for tonsil resection. The aim is to achieve uncontroversial anatomical decompression.
Results We operated 117 children during the last 10 years. Seventeen had complications, mostly hydraulic and minor; although most resolved without consequences, one patient died of unexplained cerebral vasospasm. At last control, 97% were clinically improved. No patient required reoperation for PFD.
Conclusion With proper patient selection, extensive PFD is a very efficient operation.

Keywords Chiari malformation · Child · Outcome

Introduction

Chiari malformation is clearly a misnomer. Although all medical professionals know about it, this name, since its very introduction at the end of the nineteenth century, has blurred the issue and delayed getting a clear view on the topic. We have finally come to terms with the christening quarrel initiated by Arnold's followers, the initial classification, including different types (2 to 4) which we now know are really different diseases, and the semi-quantitative approach leading to the creation of Chiari 0 and Chiari ½ subtypes. However, we are still confused by the respective roles played by intracranial hypertension, insufficient growth of the posterior calvaria, excessive growth of the cerebellum, and a myriad of rare conditions associated with this otherwise common finding. Perhaps things would get any clearer if we could achieve a consensus on:

- The correct name should be chronic tonsillar herniation (CTH).
- The anomaly is developmental rather than congenital.
- Its mechanism is mostly a hydrodynamic process rather than a malformation.
- It may evolve in time, with self-resolution as well as self-aggravation possible outcomes.

These points being made would help for decision-making and patient management.

What symptoms are considered typical?

CTH is probably the most common incidental findings in pediatric neurosurgery, even more common than

✉ Matthieu Vinchon
matthieu.vinchon@chru-lille.fr

[1] Department of Pediatric Neurosurgery, Hôpital Roger Salengro, Lille University Hospital, CHRU de Lille, 5037 Lille Cedex, France

[2] Lille University Hospital, Lille, France

Table 1 Clinical presentation

Clinical feature	Suggestive	Compatible	Differential diagnosis
Headache	Occipital, brief, exertional	Diffuse, during activity	Migraine, abuse of painkillers, ophthalmic
Sleep disturbance	Central apneas > 6/h	Awakening/pain	Obstructive apneas
Malaise	Respiratory arrest	Caused by pain asthenia related with sleep deprivation	Vagal
Neurological findings	Suspended syndrome Micronystagmus Horner's sign	Pyramidal syndrome	Cerebral palsy
Spine	Scoliosis Lt-sided, painful, in boy	Torticollis/neck pain	Persistent ependymal canal
Cognition	Learning difficulties caused by sleep disturbance	Delay caused by birth injury epilepsy caused by sleep deprivation	Incidental finding of Chiari

arachnoid cysts. When faced with this diagnosis, the question is twofold:

- Can CTH really be the cause of the symptoms, which motivated the imaging?
- If not, are we certain that the patient has really no symptom of CTH?

These questions can be difficult to answer especially in young children. Although CTH can present with a variety of signs and symptoms reflecting the protean role of the brainstem, including hallucinations and feeding problems; the most common clinical presentation for CTH is pain, sleep difficulties, neurological signs and symptoms, and signs of syringomyelia. According to the characteristics listed in Table 1, these features will be considered suggestive, compatible, or ascribed to a differential diagnosis.

The prevalence of these symptoms, compatible or typical, in our series, is shown in Fig. 1.

CTH found in young children explored for developmental delay is generally considered incidental. However, as shown below, developmental outcome can improve after surgery, probably because chronic pain and poor sleep hinder normal progress and schooling. Alternately, delay can result from birth difficulties, which can also be the cause of the CTH; in this case, CTH does not cause delay, but both result from a common cause.

Malaise is also problematic because it is extremely common and mostly benign; however, it can in rare cases herald severe brainstem malfunction inciting to rapid surgery. This highlights the fact that clinical analysis alone is not sufficient to make a decision.

Criteria for surgery

Decisions are based on clinical and paraclinical findings and must exclude differential diagnoses.

Ideally, the child presents with symptoms considered typical as detailed above, which impact enough on daily life to justify surgery. As stated above, surgery may need to be made in emergency in case of malaise caused by brainstem dysfunction.

MRI of the neuraxis shows tonsillar ectopia, thinning, effacement of arachnoid spaces, and deformation and edema of the medulla (Fig. 2). Significant syrinx should be distinguished from persistent ependymal canal. Other CSF hydrodynamic disturbances like pseudotumor, often related to obesity, and hydrocephalus should also be eliminated before deciding posterior fossa decompression (PFD).

Despite its worth, MRI can both over- and underestimate the significance of CTH. Many children whose

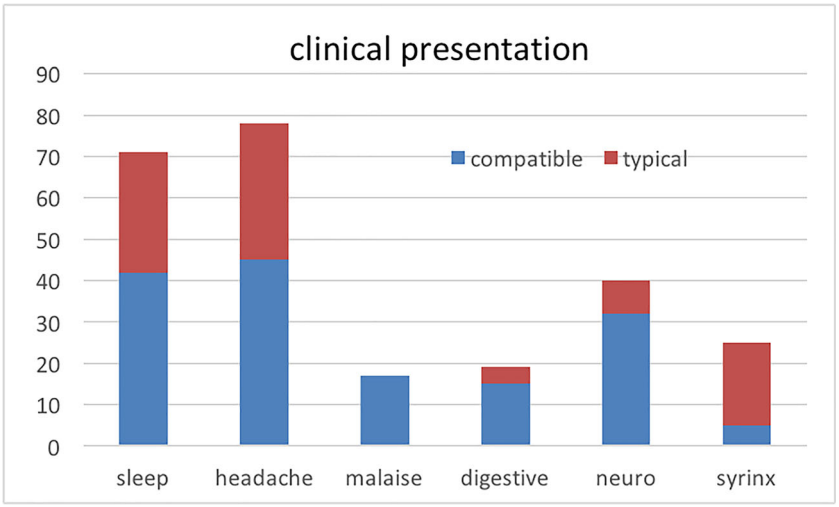

Fig. 1 Clinical presentation in our series, with typical or compatible

imaging shows impressive compression are perfectly well; some of them eventually show spontaneous regression of CTH (Fig. 3). On the other hand, MRI performed under anesthesia or in the awake calm state does not necessarily reflect what happens in stress conditions or during REM sleep: brain swelling may aggravate CTH, causing sleep apneas with hypercarbia, and initiating a vicious circle. The same caveat applies to intraoperative ultrasound.

Sleep studies are performed very commonly in order to evaluate the impact of the CTH and provide very useful, objective, and quantitative data on the significance of CTH.

In severe and non-controversial cases, however, surgery should not be delayed; in our opinion, sleep studies are especially useful in mild cases, in order not to miss a radiologically unimpressive, but clinically significant CTH.

The use of CT scanner is not systematic, but should probably become more widespread, in order to eliminate a malformation of the craniocervical junction or craniosynostosis.

Type of surgery

In our institution, we stick to an invasive approach with systematic dural opening and duraplasty (Fig. 4), the options are detailed in Table 2.

When the result is considered good: criteria for postoperative evaluation

Since PFD for CTH is mostly a functional surgery, the main evaluation criterion should be functional improvement, regarding the same items as listed preoperatively. This opens the door to biases, like parents or children unwilling to undergo another operation and declaring clinical improvement or self-complacent surgeons being easily satisfied. These biases are mitigated by the multidisciplinary follow-up in many of these patients.

In our opinion, the clinical criterion appears sufficient in the majority of cases, and we do not perform systematic postoperative imaging. We control sleep studies postoperatively only if the patient is not improved.

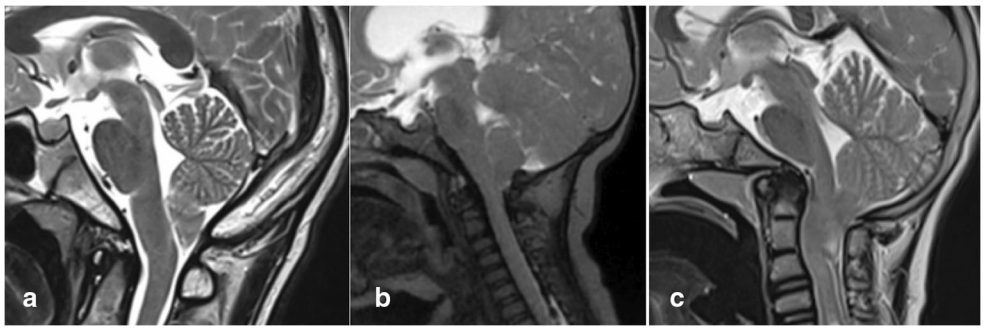

Fig. 2 T2 MR, sagittal view, showing **a** mild, **b** clear, and **c** severe tonsillar herniation. Patient A had a syrinx. All patients had significant compression found at surgery and clinical improvement postoperatively

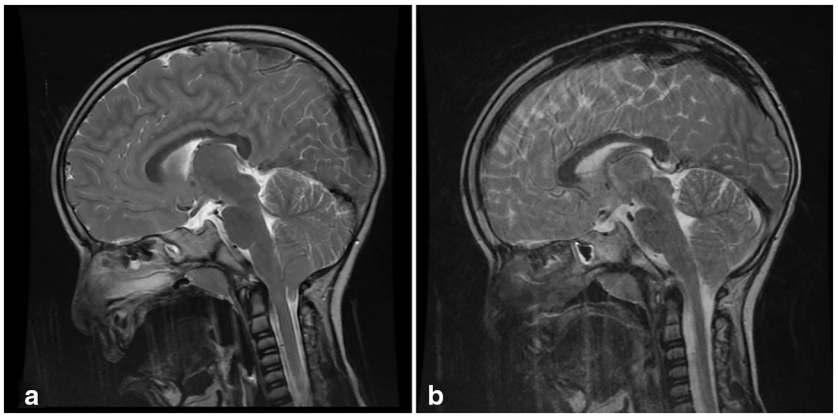

Fig. 3 **a** MRI at age 5 years, incidental finding after head trauma; **b** spontaneous regression at age 8 years

Clinical series

We reviewed children who underwent PFD for CTH in our institution during the last 10 years. Patients with occipital stenosis and complex craniocervical malformation; cases of CTH secondary to craniosynostosis were not included when the craniosynostosis required surgical management per se.

We selected 117 cases aged 8 months to 18 years (median 7.4 years), the M/F ratio is 1.39. Perinatal dif-

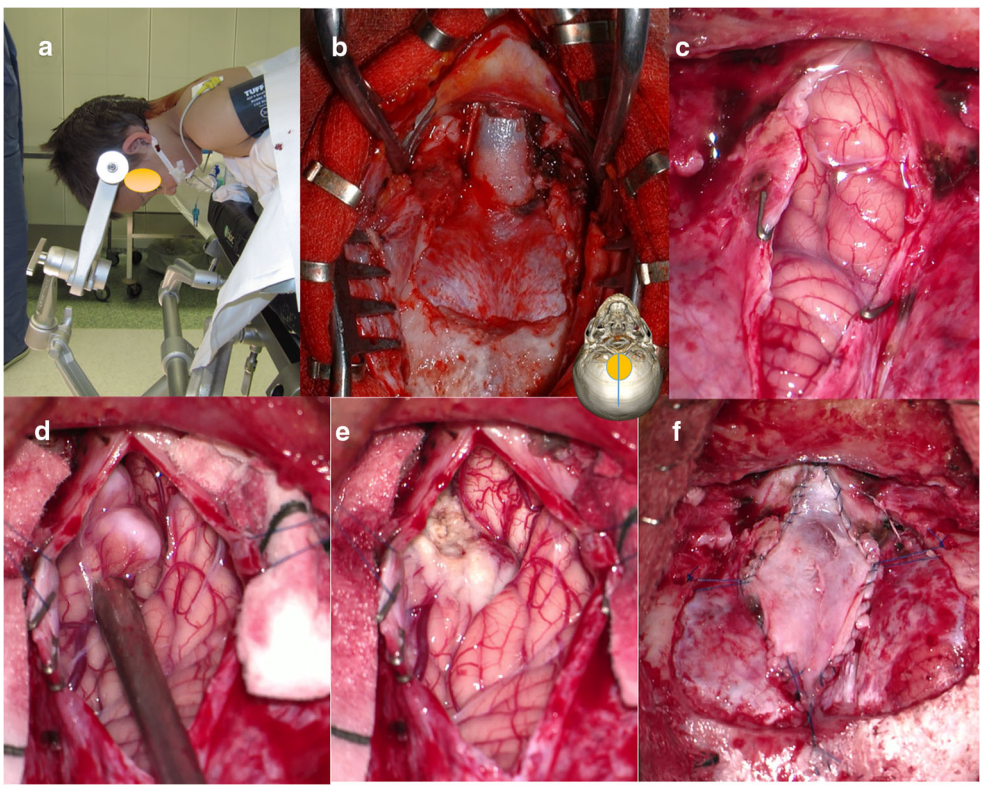

Fig. 4 Surgical technique: **a** ventral position on vacuum mattress, head flexed on pin headrest; **b** craniectomy exposing the lower posterior fossa and opening widely the foramen magnum with resection of the posterior arch of C1; **c** paramedian vertical opening of the dura mater; **d** arachnoid dissection of the tonsil (note gliosis); **e** tonsil resected; **f** duraplasty with pericranium and peripheral suspensions

Table 2 Surgical options and rationale

	Chosen option	Rationale
Bone	Craniectomy of lower posterior fossa	Create neo-cisterna magna
	Wide opening of foramen magnum	Avoid swan-neck
	Resection of posterior arch of C1	
	Respect of muscular insertions on C2	
Dura	Paramedian linear opening	Enough to expose the tonsils and obex
Tonsil	Resection if obstructive (low threshold)	Achieve unquestionable decompression
Graft	Autograft with pericranium	Autologous material
Closure	Watertight closure	Avoid meningocele
	Suspended sutures	Create neo-cisterna magna
		Avoid epidural hematoma

ficulties like dystocia or prematurity were identified in 41 (35%). The most common clinical presentations were headache and sleep problems; the prevalence of these, and other symptoms, and their interpretation as suggestive or compatible with CTH are detailed in Fig. 1.

Significant syringomyelia was found in 35 (30%), and two patients had successful but insufficient CSF drainage for hydrocephalus before undergoing PFD.

All patients underwent dural opening with autologous duraplasty; in addition, 92 (79% underwent resection of one tonsil. A ventriculo-cisternal catheter was put in place in 14 cases (12%).

One patient died after surgery of unexplained vasospasm of the polygon of Willis. Overall, 17 patients (14.5%) had complications after surgery: 15 of these were CSF-related, 2 were postoperative bleeding, and 2 required craniotomy for craniosynostosis. Overall, although 5 patients required additional surgery, none required redo surgery for PFD.

The mean follow-up was 24 months; 109 patients (97%) were clinically improved after surgery. The improvement of preoperative symptoms is detailed in Fig. 5.

Discussion

The aim of surgery for CTH is to decompress structures and restore durably normal hydrodynamics at the craniocervical junction. The surgical technique for PFD is a notoriously divisive topic, and most teams make their surgical choice based on tradition rather than scientific evidence. In our experience with extensive

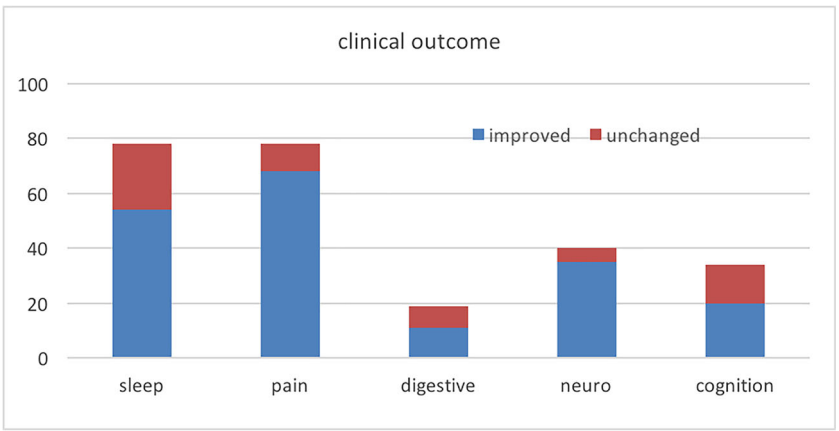

Fig. 5 Improvement in the different signs and symptoms present preoperatively

surgery, except for one unexplained death, most children were clinically improved and none required redo PFD.

Since CTH is a common chance finding, clinical and radiological findings must be scrutinized in order to decide whether surgery can benefit the patient. Determining what preoperative criteria are accurate, and whether the clinical findings are suggestive, merely compatible, or differential diagnosis, would represent an advance. This semi-quantitative approach in order to reduce uncertainty is akin to script concordance tests. It could lead to establishing a cumulative score, then setting a threshold for surgery. These preoperative criteria would then de facto be used as a benchmark for postoperative evaluation.

Compliance with ethical standards

Conflict of interest The author declares that there are no conflicts of interest.

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