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RESEARCH

Undiagnosed sagittal synostosis as cause of "idiopathic" intracranial hypertension

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Abstract

Purpose Idiopathic intracranial hypertension (IIH) is a rare condition in children, but if diagnosed needs to be promptly treated to avoid clinical sequalae. The main purpose of this paper was to test our clinical experience with a cohort of normocephalic children with craniosynostosis who do not present in the routine way to craniofacial services, due to the normal head shape and age, diagnosed with IIH.

Methods We retrospectively reviewed all children who were referred to neurosurgery from 2012 to 2022 for management of IIH on our prospectively kept database. We determined what treatments were offered and if there was an associated craniosynostosis.

Results In total, 19 children were identified with an average age at referral of 11.5 years (st dev 4.0 years) with 11 male and 8 female. The most common presenting symptoms and signs were papilloedema (18/19), headaches (15/19), visual deterioration (9/19), nausea and vomiting (7/19) and diplopia (4/19). Five out of 19 children (26.3%) had a sagittal suture fused that was not identified at the time of treatment and all children were normocephalic.

Conclusion There is a cohort of children with IIH who will have concomitant craniosynostosis and ideally would benefit from cranial vault expansion as primary surgery rather than cerebrospinal fluid (CSF) diversion. We suggest all children with IIH requiring neurosurgical intervention have cross-sectional imaging to look for occult craniosynostosis prior to intervention.

Keywords Idiopathic intracranial hypertension · Craniosynostosis · Calvarial vault expansion · Papilloedema

Introduction

Idiopathic intracranial hypertension (IIH) is a rare condition in childhood with an estimated annual incidence of about 1/100,000 children ages 1–16 years old [1, 2] with the majority of affected children in adolescence rather than early childhood [2, 3]. First-line management normally involves medication including acetazolamide [4] with other options including topiramate [5], furosemide [6] and steroids [7]. In a cohort of patients, the elevated intracranial pressure (ICP) becomes medically refractory, and they develop vision-threatening papilloedema or other stigmata of elevated ICP and require surgical intervention.

The mainstay of surgical treatment has traditionally involved cerebrospinal fluid diversion procedures in the form of lumboperitoneal shunt (LPS) or ventriculoperitoneal shunt (VPS) [5, 8]; although, other options such as optic nerve sheath fenestration [9] and venous sinus stenting [10] have also been used in children with variable success. Some consider cranial vault expansion (CVE) or decompression a "last resort" [11] surgery for IIH.

More recently we have discovered that there appears to be a cohort of normocephalic patients with undiagnosed sagittal synostosis and secondary stigmata of raised ICP [12] such as secondary IIH. This is important to recognise as we would recommend children with craniosynostosis and secondary raised ICP (or IIH as it may be mislabelled) to undergo CVE rather than CSF diversion as the first-line treatment.

The aim of this study was to evaluate the rate of undiagnosed craniosynostosis in children referred to our unit for surgical management of IIH.

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To our knowledge, this phenomenon—normocephalic children and particularly teenagers with IIH and undiagnosed craniosynostosis—has not previously been reported.

Methods

All children (under 16 years of age) with a diagnosis of IIH who were referred to our neurosurgery department in the period January 2012—January 2022 were identified using our prospectively kept database. Baseline demographic information was obtained, as well as presenting symptoms, ophthalmology findings, neurosurgical intervention, complications, outcome data and whether craniosynostosis was present or not. We also noted whether craniosynostosis was mentioned in any of the radiology reports.

Multidisciplinary approach

The management of children with IIH in our region is run in a multidisciplinary (MDT) way. General paediatricians together with paediatric neurologists manage children with

Table 1 Patient demographics and clinical information

Patient number	Age/gender	Presenting symptoms	Ophthalmology findings	Craniosynostosis/ suture	Cephalic Index	Medical management	Surgical management	Papilloedema resolved
1	7/M	Headaches, vomiting	Papilloedema	N/A		Acetozolamide	CVE	Yes
2	16/F	Visual loss	Papilloedema, visual loss	N/A			VPS	Yes
3	17/F	Headaches, nausea, vomiting	Papilloedema	N/A		Furosemide	VPS	Yes
4	15/M	Headaches	Papilloedema, visual loss	N/A		Furosemide, acetozolamide, lp	VPS	Yes
5	11/F	Visual loss, diplopia	Papilloedema, visual loss	Yes/sagittal	0.83		ICP, LPS	Yes
6	14/M	Headaches, visual loss, nausea	Papilloedema, enlarged blind spot	N/A		Acetozolamide, topiramate, lp	N/A	Yes
7	10/M	Headaches, diplopia, visual loss	CN6 palsy	N/A		Acetozolamide	ICP, VPS	Yes
8	4/F	Headaches	Papilloedema	N/A			VPS	Yes
9	12/M	Headaches	Papilloedema	Yes/sagittal	0.79		LPS, VPS, CVE	Yes
10	15/F	Headaches, vomiting	Papilloedema	N/A		Amitriptylline	VPS	Yes
11	14/M	Headaches	Papilloedema	N/A		Acetozolamide, LP	N/A	Yes
12	15/F	Headaches, visual loss	Papilloedema	N/A		Acetozolamide, LP	N/A	Yes
13	10/F	Visual loss	Papilloedema	N/A			ICP, optic nerve sheath fenestration	Yes
14	11/M	Diplopia and squint	Papilloedema	N/A		Acetozolamide	LPS	Yes
15	11/M	Headaches	papilloedema	N/A		Gabapentin	VPS	Yes
16	16/F	Headaches, visual loss, diplopia, squint	Papilloedema	N/A			VPS	Yes
17	8/M	Headaches	Papilloedema	Yes/sagittal	0.84		LPS	Yes
18	10/M	Headaches, vomiting	papilloedema	Yes/sagittal	0.76	Acetazolamide	n/a	Resolving
19	3/M	Headaches	Papilloedema, visual loss	Yes/sagittal	0.81	Acetozolamide	CVE	No

LP lumbar puncture, *LPS* lumboperitoneal shunt, *VPS* ventriculoperitoneal shunt, *ICP* Intracranial pressure bolt monitoring, *M* male, *F* female, *CVE* cranial vault expansion

a diagnosis of IIH including the workup and initial medical therapy. Unofficial MDT will often take place between paediatric neurologists and neurosurgeons regarding children unresponsive to medical treatment (consisting of serial lumbar punctures and trials of acetazolamide and other medical therapies) without a formal referral taking place. Only children truly refractory to medical management including one "therapeutic" or serial lumbar puncture with vision-threatening papilloedema are referred to a neurosurgeon formally for surgical treatment. The majority of children with IIH will generally not require surgical intervention [13].

This cohort therefore only includes children who were formally referred to neurosurgery for severe medically refractory raised ICP in diagnostically confirmed intracranial hypertension. All children in this series had a definite IIH (according to Friedman criteria) with an opening pressure (OP) > 25 cm H2O, and 18 out of 19 are also associated with papilledema. A neuro-ophthalmological evaluation was performed in all cases by a senior consultant ophthalmologist.

Results

Overall, 19 children were identified with a mean age of 11.5 years (S.D 4.0 years, range 3.2–15.6 years). There were 11 males and 8 females. See Table 1 for demographic and clinical information.

Presenting symptoms and signs

The most common presenting symptom was headache in 15/19, visual blurring/loss in 9/19, nausea and vomiting in 7/19 and diplopia in 4/19. Of the presenting signs, the most common was papilloedema in 18/19 and sixth nerve palsy in 4/19. One child without papilloedema presented with a sixth nerve palsy.

Treatment

The most common treatment offered was VPS (9/19) followed by LPS (4/19), CVE (3/19) and optic nerve sheath fenestration (1/19); one child had an LP shunt followed by a VP shunt and then subsequent CVE. Four children did not require any neurosurgical intervention ultimately and were managed with serial lumbar punctures after a formal neurosurgical review. Two further children (not included in this series) are currently on the waiting list for CVE after having been diagnosed with incidental sagittal craniosynostosis and IIH.

Craniosynostosis

Five children (5/19) were retrospectively diagnosed with craniosynostosis that had not been recognised. This was diagnosed on a computed tomography (CT) head scan (Fig. 1). The affected suture was sagittal in all cases. The mean cephalic index for these children was 0.81 with no children tending towards a scaphocephalic head. Of these, 2 children had LPS, 1 child had an LPS followed by a VPS and then a subsequent CVE, and 1 patient underwent direct CVE as the first procedure (Figs. 2 and 3) as his sagittal synostosis was diagnosed pre-operatively; the last patient has been successfully managed conservatively with acetazolamide (for 2 months) with complete resolution of the papilloedema and no surgery so far.

Complications

There were no serious complications associated with any of the neurosurgical procedures. VPS revision was required in two cases. One child was still having vision-threatening papilloedema despite the insertion of an LPS, underwent removal of LPS and insertion of VPS and ultimately, cranial vault expansion with papilloedema resolution. One child developed a secondary asymptomatic cerebellar tonsillar descent after the insertion of an LPS.

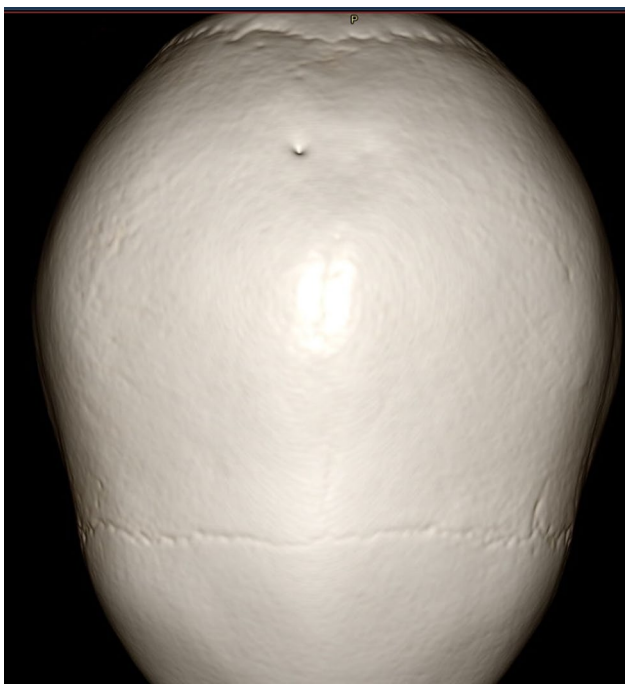

Fig. 1 Reconstructed 3D computed tomography scan from a representative case. This is a 10-year-old child who presented with headaches and papilloedema. He was normocephalic with a cephalic index of 0.76. He was diagnosed with IIH after lumbar puncture by a neurologist. He was referred to neurosurgeons for consideration of a ventriculoperitoneal shunt as he had failed an acetazolamide trial. On retrospective review of his CT head it was noted he had a sagittal craniosynostosis

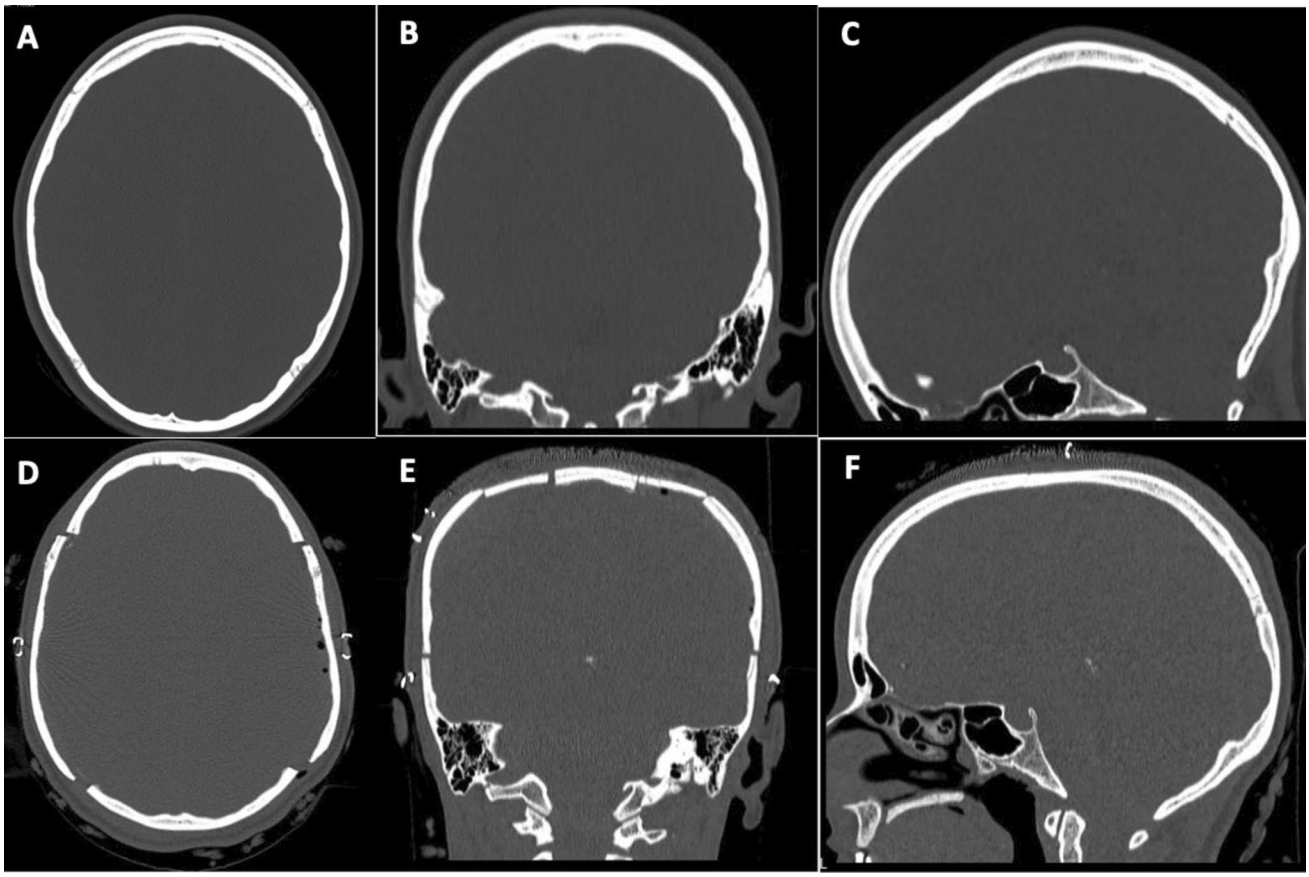

Fig. 2 CT scan (bone sequences) of patient 19 before (**A**–**C**) and after (**D**–**F**) cranial vault expansion (axial, coronal and sagittal view)

Discussion

Sagittal synostosis in normocephalic children was identified in over a quarter of patients with IIH referred for neurosurgical management. Two additional children were identified during the new diagnostic workup we have recently implemented for children with Chiari and IIH (see decision flow diagram in Fig. 4). We have planned for them a calvarial vault expansion as first-line procedure to treat what we believe to be the key driving force for the elevated intracranial pressure.

In our series, by coincidence, CVE was performed in one child with craniosynostosis and a likely secondary IIH. Had the craniosynostosis been identified earlier this would have been our preferred surgical option of choice [14, 15] in all children with craniosynostosis. Whilst LPS did work well in 2 children, it did not result in any improvement in one child with undiagnosed sagittal craniosynostosis and he required further operations before the papilloedema was resolved with a CVE (performed once the sagittal synostosis was noted on a post-operative CT scan).

Whilst we recognise that non-syndromic craniosynostosis, especially sagittal craniosynostosis, can cause a secondary raised intracranial pressure, this is mostly reported in the context of children with dolichocephaly presenting at a young age [16–18]. Here, we are discussing the epiphenomenon in a cohort of normocephalic children with an average age of 11.5 years who have presented to neurology with features of IIH. The realisation that a likely later fusion of the suture (probably after 2 years of age) and, therefore, normocephaly may lead to secondary raised ICP and subsequent sequalae has been reported by us recently in the contest of children presenting with Chiari malformation. The lack of addressing the primary driver for the intracranial elevated pressure in those cases seems to expose children to experience complications post craniovertebral decompression—such as hydrocephalus and/or CSF leakag—in a significantly higher percentage than in non-synostotic patients [12].

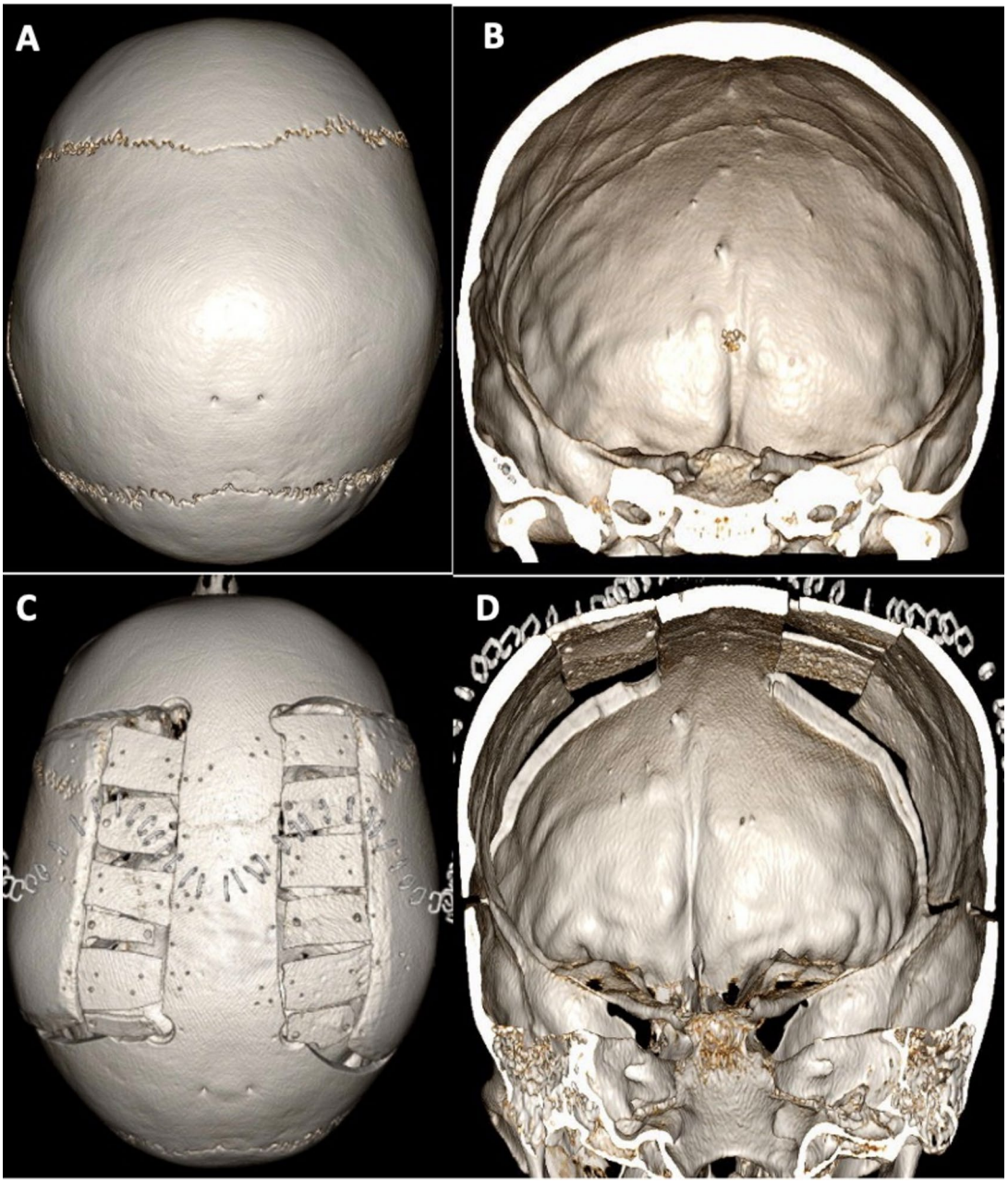

Fig. 3 3D CT scan reconstruction of patient 19 before (**A**–**B**) and after (**C**–**D**) cranial vault expansion (axial and coronal view)

This has been a learning curve for us over the years. The last two patients in this series (18 and 19) underwent a 3D CT head preoperatively as this epiphenomenon was already recognised by our team. In patient 18, acetazolamide has helped to resolve the papilloedema, and no surgery has been performed yet; in patient 19, medical treatment has been unsuccessful, and we performed a CVE as the primary procedure.

Our recommendation is that all children, including normocephalic ones, with a diagnosis of IIH should be evaluated for craniosynostosis before any surgery is planned (Fig. 4). In many cases, children will have a CT venogram to assess the state of the venous sinuses—in these cases, suture fusion can easily be seen. If a CT venogram is not available, a 3D CT scan head should be performed (or alternatively, a "black bone" MRI sequence) [16] to identify this prior to definitive neurosurgical intervention.

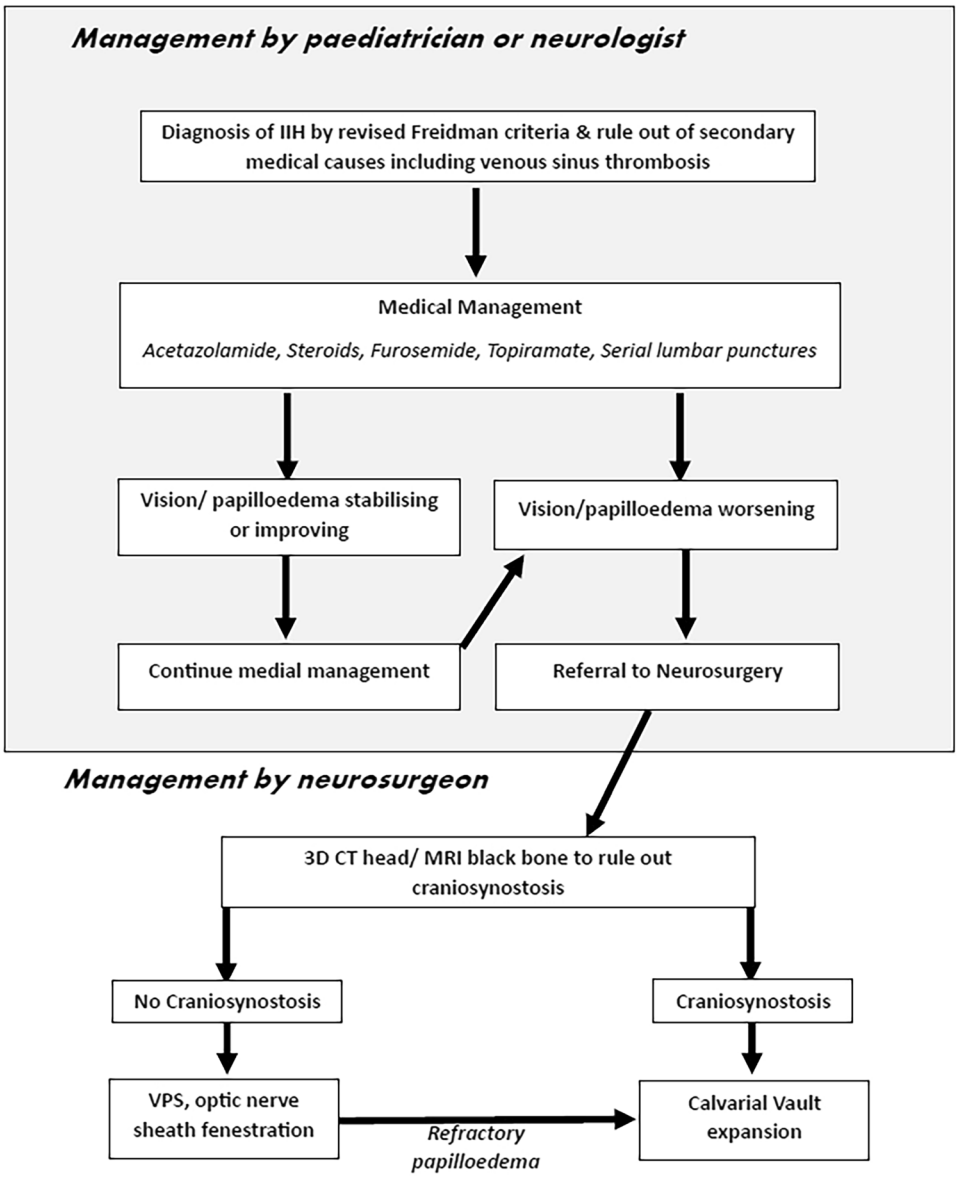

Fig. 4 Decision flow diagram for the diagnosis and surgical management of IIH secondary to craniosynostosis

Conclusions

This study has revealed that more than one in four normocephalic children referred for neurosurgical treatment of IIH had an underlying undiagnosed sagittal synostosis. Although this high incidence needs to be confirmed by other larger, and possibly multicentre series, we think it is crucial to distinguish these patients from the true idiopathic IIH to avoid unnecessary and/or potentially harmful surgical treatments.

Author contributions J.P. drafted the manuscript and prepared Table 1, Figs. 1 and 4. S.B. helped with data collection and analysis, P.G. designed the study and contributed to the manuscript drafting. Prepared Figs. 2 and 3. Revised the manuscript. All authors reviewed the final rmanuscript.

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Data availability No datasets were generated or analyzed during the current study.

Declarations

Conflict of interest Authors report no competing or conflict of interests.

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