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CASE-BASED UPDATE

# Conservative and operative management of iatrogenic craniocerebral disproportion—a case-based review

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## Abstract

**Introduction** Craniocerebral disproportion (CCD) can occur as a sequela after shunting in early infancy. It can be understood as a disorder closely related to slit ventricle syndrome and chronic overdrainage syndrome. Here, we present two exemplary cases and summarize the pathophysiological, diagnostic, and therapeutic approaches to CCD.

**Clinical presentation** Two premature babies underwent shunting for posthemorrhagic hydrocephalus and presented in later childhood with recurrent episodes of symptomatic raised intracranial pressure (ICP) at 2 and 8 years of age, respectively.

**Diagnosis and management** Both patients had unchanged ventricular size on cranial imaging and fulfilled the clinical diagnostic criteria of CCD. After confirming shunt patency, ICP monitoring was performed to diagnose intermittent intracranial hypertension. Different treatment pathways were pursued: While readjustment of a programmable shunt valve was sufficient to alleviate the raised ICP in the first case, a cranial expansion surgery was necessary in the second case.

**Outcome and conclusions** Both children were treated successfully after thorough assessment and careful choice of treatment approaches. This review provides detailed insight into CCD and highlights the importance of individual and critical decision-making in these complex patients.

**Keywords** Intracranial hypertension · Shunt · Chronic overdrainage · Slit ventricles

## Introduction

Craniocerebral disproportion (CCD) has been defined as a state where the volume of the brain exceeds the intracranial space, leading to inadequate accommodation of the growing brain or of physiological volume fluctuations, such as cerebral blood volume [1]. The secondary form of CCD has been attributed to shunting in infancy or early childhood and is thus considered an iatrogenic phenomenon [2]. It shares common features with slit ventricle syndrome (SVS) and chronic overdrainage syndrome and due to inconsistent or overlapping definitions, the terms are unfortunately sometimes used as synonyms [3–5]. It is thought to occur in less than 2% of children shunted in infancy [6].

The pathophysiological concept underlying CCD can be traced back to the Monro-Kellie doctrine: The skull is considered to be a fixed inelastic container, and therefore, a volumetric increase in one of the intracranial contents will lead to a compensatory decrease in another or to an increase in intracranial pressure (ICP) [7]. In CCD, the inelastic container is too small to accommodate the growing brain or physiological volume fluctuations, such as temporary increase of cerebral blood volume, and thus raised ICP will occur. The underlying reason for the inadequately small intracranial volume in secondary CCD is shunting in infancy or early childhood (usually in the first year of life), which influences intracranial volume and cerebrospinal fluid (CSF) pulse pressure. These two parameters are considered important mechanical forces, which under physiological conditions are modulators of calvarial expansive growth, i.e., skull growth is triggered by brain growth [8].

The morphological hallmarks of CCD comprise microcephaly, thickening of the skull bone with copper-beaten

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appearance of the inner table, premature fusion of cranial sutures, slit-like ventricles, effacement of cortical subarachnoid spaces, and hindbrain herniation. Sandler et al. have hypothesized that a pressurized cortical subarachnoid space is the universal finding in CCD: The growing brain is surrounded by an inadequately small, prematurely fused, and thus inelastic skull, leading to the cascade of morphological changes of the skull, brain, and ventricular system during further development, summarized above as the hallmarks of CCD [1].

## Historical background

As the 1950s and 1960s saw significant advances in shunt technology and increasing numbers of shunt procedures being performed, literature on complications and side effects began to emerge [9]. Among the known common shunt complications, "craniosynostosis developing with the approximation of the parietes after a successful ventriculo-atriostomy" was briefly mentioned by Strenger in 1963 [10]. Case reports and small case series of similar observations in shunted patients soon followed [11, 12]. The radiological changes occurring in affected patients were formally described and in 1966, the term "hyperostosis cranii ex vacuo" was coined by Moseley et al. to distinguish thickening of the calvarium and premature closure of cranial sutures in shunted children from other forms of calvarial disorders [13]. The foundation of the current concept of CCD was laid in 1976 with the description of "cephalocranial disproportion" as a "complication of the treatment of hydrocephalus in children" by Hoffman and Tucker [14].

## Clinical presentation

CCD is clinically characterized by chronic, often incapacitating headaches in the presence of a functioning cerebrospinal fluid (CSF) shunt, which had been implanted in infancy or early childhood [1, 15]. In our experience, these headaches typically occur in episodes and, unlike pure low-pressure headache associated with CSF overdrainage, are not reliably relieved by lying flat. During episodes of raised ICP, other typical signs and symptoms of intracranial hypertension can be observed, such as lethargy or Parinaud syndrome. In children with long-standing CCD, developmental delay, decline in school performance and social withdrawal can be sequelae of chronic severe headache interfering with activities of daily life. A typical patient suffering from CCD will have a history of multiple shunt revisions, ranging from a mean number of five revisions prior to cranial expansion surgery to extreme numbers of up to 40 previous revisions in young adults [16, 17].

## Diagnosis

Measurement of head circumference and ear-to-ear distance will typically indicate below-average head size or microcephaly. The morphological hallmarks of CCD can be visualized by cranial magnetic resonance imaging (MRI) and cranial computed tomography (CT). Of note, serial cranial imaging in affected children typically remains unchanged with regard to ventricular size even during episodes of severe headache [15]. In addition to radiological imaging, shunt taps with manometric assessment are often performed as the next step to ensure shunt patency. Ophthalmological examination with regard to papilledema can be helpful, although papilledema can be absent when intracranial hypertension occurs in an episodic fashion. Finally, invasive ICP monitoring is recommended to elucidate the origin of symptoms in affected children, i.e., low or high ICP, and thus coordinate and guide further treatment [3, 18, 19]. Sandler et al. have suggested a diagnostic and therapeutic algorithm for shunted patients complaining of chronic headache, which in our experience offers excellent clinical guidance [1].

## Management

If iatrogenic CCD is suspected, the clinical algorithm suggested by Sandler et al. can provide guidance regarding diagnostic and therapeutic steps [1]. In affected children, ventricular size typically remains stable even during episodes of acute headache exacerbation. If ventricular dilatation is evident, shunt failure should be suspected. If cranial imaging remains unchanged during headache episodes, the patency of the shunt system has to be verified. In these children, repeated emergent CT imaging should be avoided as it is futile and carries a risk of radiation-induced malignancy.

If a patent shunt system can be confirmed, ICP monitoring should be performed as the next step. In our experience, implantation of a telemetric ICP probe offers the advantage of long-term monitoring even in the outpatient setting. In selected patients, an attempt to relieve headache episodes by shunt valve readjustment should be considered. In our practice, such initial adjustments are made under inpatient observation and guidance by ICP monitoring. The basic goals of treatment should be the relief from headaches and normalization of ICP during all positions and during sleep. If conservative approaches fail and episodically raised ICP still occurs during headache episodes, cranial vault expansion is indicated [1].

## Prognosis and outcomes

The best treatment of CCD in shunted patients would be its avoidance. While it has to be acknowledged that no particular shunt system can be recommended according to the criteria of evidence-based medicine, several studies at least indicate that

shunts without overdrainage protection or low-pressure shunts should be avoided especially in infancy and early childhood [20–24]. The efficacy of anti-siphon devices in general has been demonstrated also in in vitro studies [25]. Based on this data, we hypothesize that avoidance of chronic overdrainage in children shunted in infancy and early childhood would reduce the risk of subsequent iatrogenic CCD.

A systematic literature review of cranial expansion surgery for iatrogenic CCD secondary to CSF shunting identified 13 publications describing 64 patients (Table 1) [16, 17, 26–36]. The craniofacial procedures described in these publications ranged from strip craniectomies with barrel-stave osteotomies to full calvarial remodeling procedures and use of distraction devices. However, irrespective of surgical techniques, the authors achieved significant clinical improvement or complete resolution of symptoms. A regression of hindbrain herniation has been reported after supratentorial cranial expansion [37]. The complication rate appears to be low, suggesting that cranial expansion surgery for iatrogenic CCD is effective and has a favorable risk-benefit ratio. Concerning underlying pathophysiological aspects, Cornelissen et al. recently demonstrated a decreased mean venous flow velocity in the superior sagittal sinus in children with craniosynostosis [38]. After surgical intervention, an improvement of superficial venous drainage was noted.

## Exemplary case description 1—conservative approach

This male patient was born prematurely at 27 weeks of gestation and suffered intraventricular hemorrhage with consecutive posthemorrhagic hydrocephalus. After temporizing treatment with a ventricular access device, he received a ventriculoperitoneal shunt (VPS) with a fixed pressure gravitational valve (Miethke paediGAV 4/19 $cmH_2O$, B. Braun/Aesculap AG, Tuttlingen, Germany) within the first months of life. Shunt malfunctions due to ventricular catheter occlusion occurred at an age of 5 and 6 years, followed by a long period of unremarkable outpatient follow-up visits. However, he was noted to have slit ventricles on routine imaging.

At an age of 8 years, he presented repeatedly with episodes of intractable headaches and vomiting, inconsistently exacerbated in the upright position. He underwent shunt revision with switch to a Miethke paediGAV 9/19 $cmH_2O$ valve (B. Braun/Aesculap AG, Tuttlingen, Germany), followed by two further revisions for suspected shunt failure within weeks. He finally presented with persistent headache, lethargy, bradycardia, and arterial hypertension. An external ventricular drain was inserted and complete remission of symptoms was noted with 200–300 ml of CSF drainage per day, maintaining an ICP well below 10 $cmH_2O$. A programmable valve (Codman Hakim valve, Codman & Shurtleff Inc., Raynham, USA) adjusted at 60 $mmH_2O$ was implanted, combined with an anti-siphon device (Miethke Shunt-Assistant, B. Braun/Aesculap AG, Tuttlingen, Germany) with a 20 $cmH_2O$ pressure level.

He remained well for 2 years, until he again presented with similar but aggravated episodes. Cranial imaging demonstrated an unchanged ventricular size with persistent slit ventricles, copper-beaten appearance of the inner table and hindbrain herniation (Fig. 1). The shunt was patent during reservoir tap and papilledema was excluded. Head circumference was 46 cm, which is approximately 4.5 cm below the 3% percentile for this age. An ICP probe (Raumedic AG, Helmbrechts, Germany) was inserted. Prolonged ICP monitoring during headache exacerbation demonstrated highly pathological recurrent plateau waves and B-wave storms with a mean ICP of 29 mmHg at night (Fig. 2).

As an additional option to the algorithm suggested by Sandler et al., we decided to increase the programmable valve pressure level to 10 $cmH_2O$ in an attempt to reexpand the collapsed ventricles and achieve a more continuous CSF drainage [1]. This approach was successful and no further episodes of exacerbated headaches occurred, although it remains to be seen whether an operative approach to correct the overt CCD in this child can be avoided over the long-term. However, telemetric ICP monitoring during an outpatient clinic visit after 8 months demonstrated a more physiological posture-dependent ICP pattern (Fig. 3).

## Exemplary case description 2—operative approach

This female patient was born prematurely at 26 weeks of gestation and suffered intraventricular hemorrhage with consecutive posthemorrhagic hydrocephalus. After temporizing treatment with a ventricular access device, he received a ventriculoperitoneal shunt (VPS) with a fixed pressure gravitational valve (Miethke paediGAV 4/19 $cmH_2O$, B. Braun/Aesculap AG, Tuttlingen, Germany) within the first months of life.

At an age of 2 years, she presented with recurrent episodes of impaired consciousness, vomiting, and seizures. A dolichocephalic head shape was noted with a cephalic index of 63. The head circumference was 41.5 cm, which was 4 cm below the 3% percentile for this age. Serial MRI and CT imaging revealed slit ventricles with unchanged ventricular size regardless of clinical status (Fig. 4a). Two shunt revisions were performed within 6 months and a gravitational valve with higher pressure level was implanted (Miethke paediGAV 4/24 $cmH_2O$, B. Braun/Aesculap AG, Tuttlingen, Germany). However, she presented with further episodes despite unremarkable shunt tap. Invasive ICP monitoring was performed, revealing a mean ICP of 10 mmHg with scattered plateau waves vaguely corresponding to symptom exacerbation. The diagnosis of symptomatic CCD was confirmed. As she had a fixed pressure valve, no option of opening pressure readjustment was available and expansile cranioplasty was preferred

**Table 1** Summary of a systematic literature review on craniofacial surgery for iatrogenic CCD

| Author, year | *N* | Symptoms | Diagnostic approach | Treatment | Outcome |
|---|---|---|---|---|---|
| Epstein, 1988 [26] | 14 | Intermittent headache, vomiting, obtundation, bradycardia, hypertension, and extensor posturing | Shunt patency confirmed with radionuclide scan, ICP monitoring, small calvarium noted | Calvarial extension procedure | Improvement |
| Eide, 2001 [27] | 3 | Headache, nausea, vomiting, and impaired vision | Radiographic signs of CCD and slit ventricles | Anterior cranial vault expansion (1), posterior approach with modified tiara plastic (2) | Improvement |
| Martinez-Lage, 2006 [28] | 4 | Headache, drowsiness, papilledema, ptosis, and ataxia | Cranial imaging demonstrates signs of CCD, ICP monitoring | Bilateral temporo-parietal craniotomies, ± frontal advancement, and ± progressive upgrading of the programmable valve settings | Improvement |
| Weinzweig, 2008 [16] | 12 | Headache, bradycardia, papilledema, visual impairment, and seizures | Cranial imaging demonstrates (secondary) craniosynostosis | Frontoorbital advancement with frontotemporoparietal expansion; posterior vault expansion | Improvement in 8 of 12 patients |
| De Lima, 2013 [29] | 2 | Headaches, absence seizures, upgaze palsy, and lethargy | Cranial imaging demonstrates signs of SVS and CCD; ICP monitoring demonstrates raised ICP | Bilateral temporo-parieto-occipital osteotomies with internal cranial distractors | Improvement (1) and complete resolution (1) |
| Sandler, 2013 [30] | 2 | Headache, vision changes, and aggression | Shunt patency confirmed with nuclear shuntogram, unchanged ventricular size, and ICP monitoring | Bifrontal craniotomy with internal distraction devices | Complete resolution |
| Ryoo, 2014 [31] | 1 | Lethargy, seizures, and vomiting | Radiographic signs of slit ventricle syndrome combined with secondary sagittal craniosynostosis | Barrel-stave osteotomies and sagittal strip craniectomy | Complete resolution |
| Beuriat, 2015 [32] | 5 | Headache, developmental delay, impaired vision, and seizures | Cranial imaging demonstrates signs of SVS and CCD; ICP monitoring demonstrates raised ICP | Splitting bifrontal bone flap that is left floating posteriorly | Improvement |
| Bhadkamkar, 2015 [33] | 1 | Headache, developmental delay | Cranial imaging demonstrates turribrachycephalic head shape, small ventricles | Bilateral parieto-occipital craniotomy and internal distractors | Improvement |
| Mohan, 2015 [34] | 1 | Symptoms of raised ICP, microcephaly | Radiographic signs of CCD | Barrel-stave osteotomies and strip craniectomies | Complete resolution |
| Kim, 2017 [35] | 3 | Clinical symptoms of increased intracranial pressure | Radiographic signs of CCD | Biparietal craniotomies (2) and frontoparieto-occipital reconstruction (1) and simultaneous replacement of a valve with a programmable one (2) | Improvement |
| Golinko, 2018 [36] | 13 | Chronic headaches, papilledema, seizures, or behavioral changes | Radiographic signs of CCD and secondary craniosynostosis | Frontoorbital advancement (7), total cranial vault remodeling (4), posterior cranial vault remodeling, and strip craniectomy (2) | Improvement (6 of 8 with improvement of headache) |
| Winston, 2018 [17] | 3 | Headaches, nausea, and blurred vision | Cranial imaging demonstrates signs of SVS and CCD, ICP monitoring | Bilateral frontoparietal osteotomies with cranial distractors | Complete resolution |

over a simple shunt valve exchange. She underwent a cranial morcellation procedure and implantation of a new adjustable gravitational shunt (Miethke proGAV, B. Braun/Aesculap AG, Tuttlingen, Germany) with an unremarkable perioperative course. The postoperative cephalic index had increased to 72 and an expansion of the ventricular system was evident (Fig. 4b). No further episodes of impaired consciousness, headaches, or vomiting have occurred since then.

## Discussion

Iatrogenic CCD in shunted patients is a challenging phenomenon: It inflicts significant additional morbidity on the patient and effective treatment can require invasive neurosurgical procedures. On the other hand, there is a lack of data from larger prospective studies, the definitions and diagnostic criteria are divergent, and the pathophysiological understanding as well

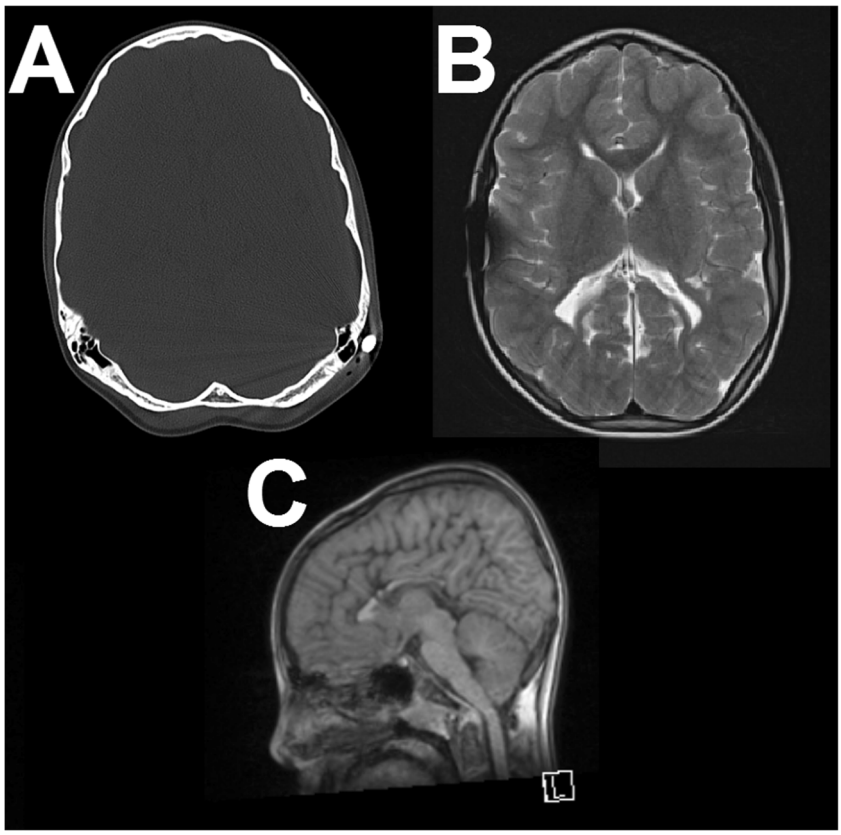


**Fig. 1** Imaging features of CCD: copper-beaten appearance of the inner table of the calvarium (**a**), small or slit ventricles (**b**), and compressed cortical subarachnoid spaces as well as hindbrain herniation (**c**)

as suggested solutions varies across the neurosurgical community.

In a recent survey among American pediatric neurosurgeons on the management of shunt-dependent hydrocephalus

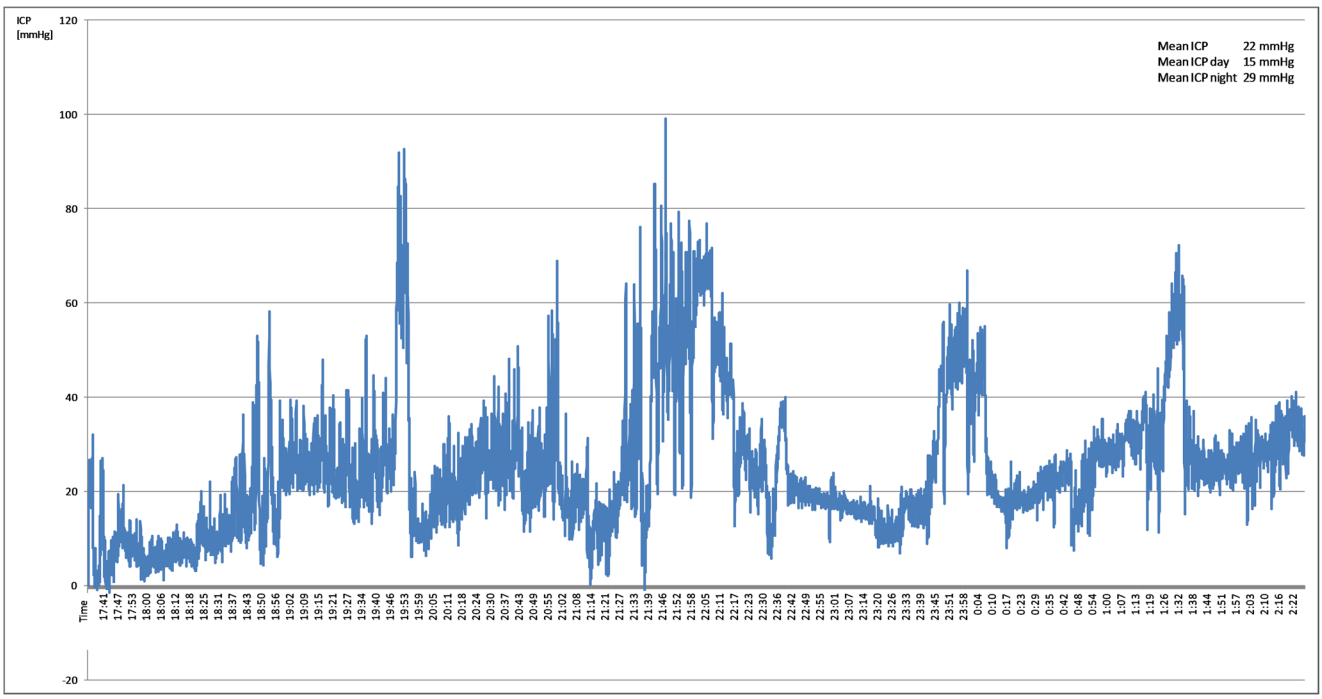


**Fig. 2** Prolonged ICP monitoring in a child with symptomatic CCD during headache exacerbation, demonstrating pathological recurrent plateau waves and B wave storms with raised mean ICP

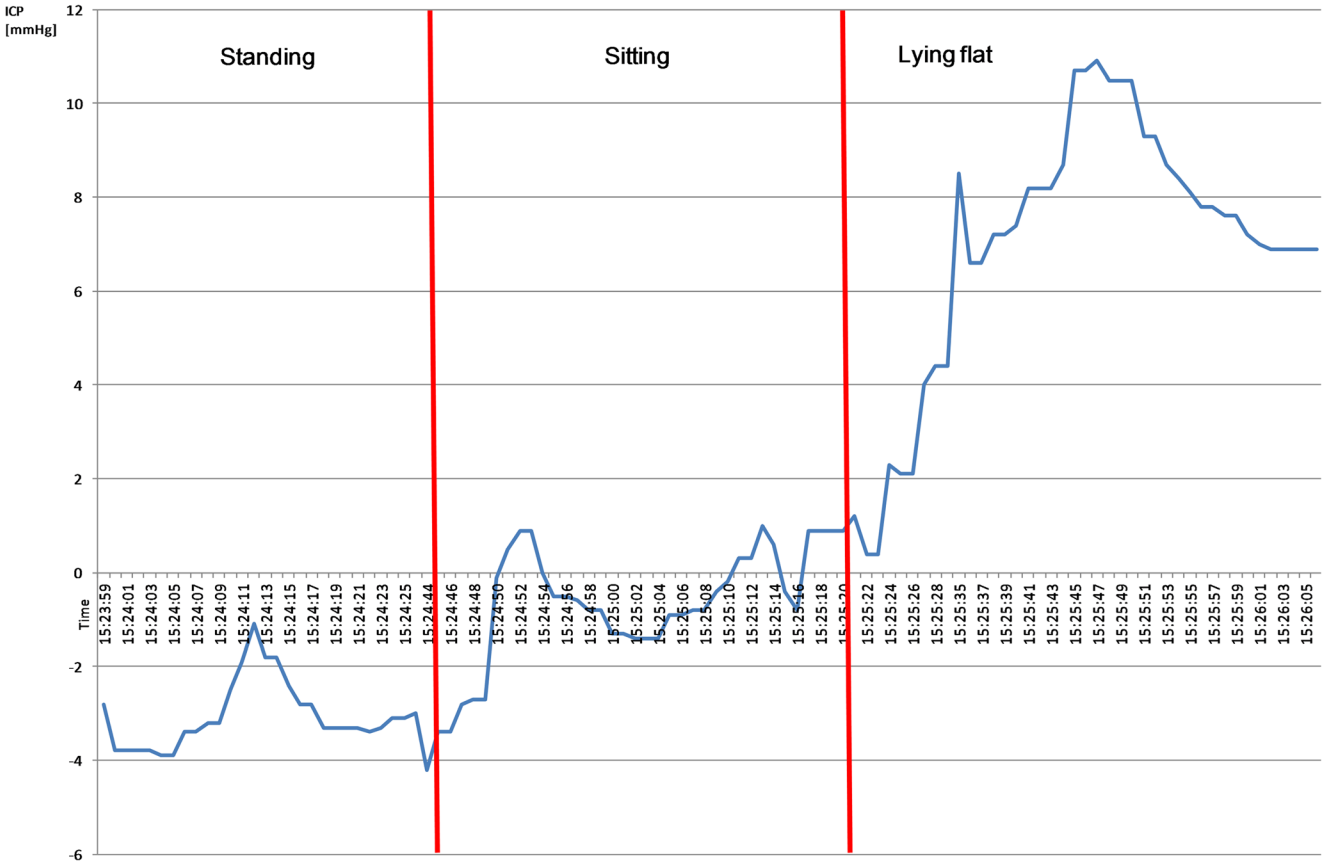


**Fig. 3** ICP monitoring of the same child as in Fig. 1 after readjustment of the shunt valve, with complete resolution of symptoms and a more physiological posture-dependent ICP pattern

published by Kraemer et al., siphoning, overdrainage, and small ventricles were commonly reported causes of ventricular catheter obstruction [39]. Chronic headaches in shunted patients were mainly attributed to medical reasons, although overdrainage was considered a more likely cause than underdrainage. For 20% of neurosurgeons, ICP monitoring played a role in the diagnostic evaluation of chronic headaches. Respondents in this survey estimated that a median of 5% of their patients suffer from chronic overdrainage, with estimates ranging between 0 and 75%. Of note, the authors mention that respondents expressed concerns about the definition of chronic overdrainage and suggested different entities: Absolute overdrainage was considered the cause of postural headaches and subdural collections, whereas relative overdrainage predisposes for SVS. In this study, SVS was defined as symptomatic intermittent ICP elevation with small ventricles. Respondents identified chronic overdrainage, abnormal brain compliance, small skull size, cerebro-venous congestion, and shunt placement in early childhood as the major causes of SVS. ICP monitoring is performed by more than 50% of neurosurgeons and shunt revision, cranial expansion surgery, and addition of an anti-siphon device are the most common treatment options. Thus, the results of this survey of actual clinical practice among pediatric neurosurgeons reflect the conceptual framework and clinical algorithm suggested by Sandler et al. quite well, although an overlap of chronic overdrainage, SVS and CCD is evident [1, 39].

In our experience, ICP monitoring is a useful diagnostic tool in investigating these three clinical entities. As an addition to the previously published algorithm, we would suggest to add shunt valve readjustments to the clinical pathway even in the presence of raised ICP. Intermittent ventricular catheter obstruction can be the underlying cause of episodes of severe headaches correlating with high ICP [15]. For chronic overdrainage syndrome, sudden venous entrapment following chronic venous distention as a result of chronic overdrainage has been suggested before as the underlying pathophysiological key mechanism, with adjustments of the operational pressure of an overdrainage preventing shunt valve being an effective treatment option [40, 41]. Typically, an increase in the operational pressures of shunt valves or gravitational units is required, which might appear counterintuitive in some cases. However, the intermittent nature of the problem has to be embraced. As we assume a close relationship or overlap between chronic overdrainage, SVS, and CCD, we applied this treatment option to children with CCD and were able to achieve clinical improvement, as demonstrated in the exemplary case one. ICP monitoring, preferentially with a

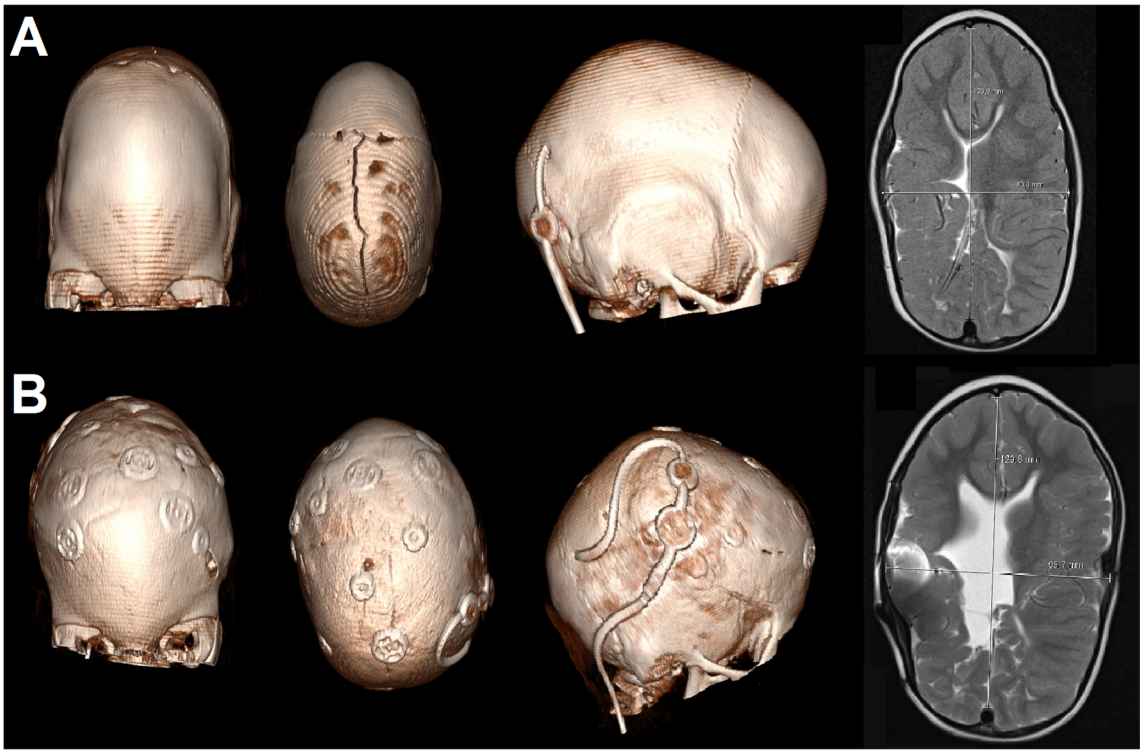


**Fig. 4** Cranial MRI of a child with CCD showing a dolichocephalic head shape and slit ventricles before surgery (**a**) and an improvement of cephalic index and ventricular size after cranial expansion surgery (**b**)

telemetric system that can be used over a long period of time, and adjustable valve with overdrainage protection are thus the keys to this treatment option [42, 43].

As a final remark, we would like to emphasize that microcephaly diagnosed by head circumference measurement alone does not by itself imply a pathological situation that requires neurosurgical intervention. From a technical point of view, the diagnostic value of head circumference measurement is limited and it is prone to measurement errors [44, 45]. Its value can be increased by additional measurement of ear-to-ear distance [44]. Hydrocephalic children shunted in infancy are known to have smaller head circumference compared to healthy children, but they also have shorter height and lower weight than healthy peers [6]. From a pathophysiological perspective, we can extrapolate data from children with craniosynostosis, where volume measurement alone is not a reliable predictor of raised ICP [46]. From a clinical point of view, extreme head size is neither specific nor sensitive for neurocognitive impairment and the severity of the underlying primary pathology, such as grade of intraventricular hemorrhage of prematurity, appears to be the major determinant of outcome [45, 47].

## Conclusions

CCD is defined as a state where the volume of the brain exceeds the intracranial space, leading to inadequate accommodation of the growing brain or of physiological volume fluctuations. The secondary form of CCD is an iatrogenic phenomenon occurring in less than 2% of children shunted in infancy. It shares common features with slit ventricle syndrome (SVS) and chronic overdrainage syndrome, suggesting prophylactic avoidance of shunts without overdrainage protection or low-pressure shunts. The diagnosis of CCD should be established following a diagnostic algorithm including assessment of the shunt, radiological imaging, and ICP monitoring. While readjustment of a programmable shunt valve can be helpful in selected cases, a subset of children will require cranial expansion surgery with a favorable risk-benefit ratio.

## Compliance with ethical standards

**Conflict of interest** On behalf of all authors, the corresponding author states that there is no conflict of interest.

## References

1. Sandler AL, Goodrich JT, Daniels LB, Biswas A, Abbott R (2013) Craniocerebral disproportion: a topical review and proposal toward a new definition, diagnosis, and treatment protocol. Childs Nerv Syst 29:1997–2010. https://doi.org/10.1007/s00381-013-2257-7
2. Di Rocco C, Frassanito P (2015) Cranio-cerebral disproportion as a late complication. In: Di Rocco C, Turgut DM, Jallo G, Martínez-Lage DJF (Eds.) Complications of CSF shunting in hydrocephalus -

prevention, identification, and management, 1st edn., Springer International Publishing, Switzerland, pp 233-246
3. Rekate HL (1993) Classification of slit-ventricle syndromes using intracranial pressure monitoring. Pediatr Neurosurg 19:15–20. https://doi.org/10.1159/000120694
4. Rekate HL (2004) The slit ventricle syndrome: advances based on technology and understanding. Pediatr Neurosurg 40:259–263. https://doi.org/10.1159/000083737
5. Di Rocco C (1994) Is the slit ventricle syndrome always a slit ventricle syndrome? Childs Nerv Syst 10:49–58
6. Nilsson D, Svensson J, Korkmaz BA, Nelvig H, Tisell M (2013) Decreased head circumference in shunt-treated compared with healthy children. J Neurosurg Pediatr 12:483–490. https://doi.org/10.3171/2013.8.PEDS1370
7. Wilson MH (2016) Monro-Kellie 2.0: the dynamic vascular and venous pathophysiological components of intracranial pressure. J Cereb Blood Flow Metab 36:1338–1350. https://doi.org/10.1177/0271678X16648711
8. Moss ML (1975) Functional anatomy of cranial synostosis. Childs Brain 1:22–33
9. Aschoff A, Kremer P, Hashemi B, Kunze S (1999) The scientific history of hydrocephalus and its treatment. Neurosurg Rev 22:67–93. https://doi.org/10.1007/s101430050035
10. Strenger L (1963) Complications of ventriculovenous shunts. J Neurosurg 20:219–224. https://doi.org/10.3171/jns.1963.20.3.0219
11. Andersson H (1966) Craniosynostosis as a complication after operation for hydrocephalus. Acta Paediatr 55:192–196. https://doi.org/10.1111/j.1651-2227.1966.tb15225.x
12. Kloss JL (1968) Craniosynostosis secondary to ventriculoatrial shunt. Arch Pediatr Adolesc Med 116:315. https://doi.org/10.1001/archpedi.1968.02100020317015
13. Moseley JE, Rabinowitz JG, Dziadiw R (1966) Hyperostosis cranii ex vacuo. Radiology 87:1105–1107. https://doi.org/10.1148/87.6.1105
14. Hoffman HJ, Tucker WS (1976) Cephalocranial disproportion. A complication of the treatment of hydrocephalus in children. Childs Brain 2:167–176
15. Rekate HL (2008) Shunt-related headaches: the slit ventricle syndromes. Childs Nerv Syst 24:423–430. https://doi.org/10.1007/s00381-008-0579-7
16. Weinzweig J, Bartlett SP, Chen JC, Losee J, Sutton L, Duhaime AC, Whitaker LA (2008) Cranial vault expansion in the management of postshunt craniosynostosis and slit ventricle syndrome. Plast Reconstr Surg 122:1171–1180. https://doi.org/10.1097/PRS.0b013e3181858c84
17. Winston KR, French B, Bunn J (2018) Chronic debilitating headache in adults caused by craniocerebral disproportion : treatment by cranial vault expansion clinical material. https://doi.org/10.7759/cureus.2187
18. Sæhle T, Eide PK (2015) Intracranial pressure monitoring in pediatric and adult patients with hydrocephalus and tentative shunt failure: a single-center experience over 10 years in 146 patients. J Neurosurg 122:1076–1086. https://doi.org/10.3171/2014.12.JNS141029
19. Tamburrini G, Caldarelli M, Massimi L, Santini P, di Rocco C (2005) Intracranial pressure monitoring in children with single suture and complex craniosynostosis: a review. Childs Nerv Syst 21:913–921. https://doi.org/10.1007/s00381-004-1117-x
20. Tschan CA, Antes S, Huthmann A, Vulcu S, Oertel J, Wagner W (2014) Overcoming CSF overdrainage with the adjustable gravitational valve proSA. Acta Neurochir 156:767–776; discussion 776. https://doi.org/10.1007/s00701-013-1934-3
21. Lemcke J, Meier U, Müller C et al (2013) Safety and efficacy of gravitational shunt valves in patients with idiopathic normal pressure hydrocephalus: a pragmatic, randomised, open label, multicentre trial (SVASONA). J Neurol Neurosurg Psychiatry 84:850–857. https://doi.org/10.1136/jnnp-2012-303936
22. Weinzierl MR, Hans F-J, Stoffel M, Oertel MF, Korinth MC (2012) Experience with a gravitational valve in the management of symptomatic overdrainage in children with shunts. J Neurosurg Pediatr 9:468–472. https://doi.org/10.3171/2012.1.PEDS11110
23. Gebert AF, Schulz M, Haberl H, Thomale U-W (2013) Adjustments in gravitational valves for the treatment of childhood hydrocephalus-a retrospective survey. Childs Nerv Syst 29:2019–2025. https://doi.org/10.1007/s00381-013-2160-2
24. Thomale U-W, Gebert AF, Haberl H, Schulz M (2013) Shunt survival rates by using the adjustable differential pressure valve combined with a gravitational unit (proGAV) in pediatric neurosurgery. Childs Nerv Syst 29:425–431. https://doi.org/10.1007/s00381-012-1956-9
25. Gehlen M, Eklund A, Kurtcuoglu V, Malm J, Schmid Daners M (2017) Comparison of anti-siphon devices-how do they affect CSF dynamics in supine and upright posture? Acta Neurochir 159:1389–1397. https://doi.org/10.1007/s00701-017-3249-2
26. Epstein F, Lapras C, Wisoff JH (1988) "Slit-ventricle syndrome": etiology and treatment. Pediatr Neurosurg 14:5–10. https://doi.org/10.1159/000120354
27. Eide PK, Helseth E, Due-Tønnessen B, Lundar T (2001) Changes in intracranial pressure after calvarial expansion surgery in children with slit ventricle syndrome. Pediatr Neurosurg 35:195–204. https://doi.org/10.1159/000050421
28. Martínez-Lage JF, Ruiz-Espejo Vilar A, Pérez-Espejo MA, Almagro MJ, Ros de San Pedro J, Felipe Murcia M (2006) Shunt-related craniocerebral disproportion: treatment with cranial vault expanding procedures. Neurosurg Rev 29:229–235. https://doi.org/10.1007/s10143-006-0022-z
29. De Lima MHN, Harshbarger RJ, George TM (2014) Treatment of cephalocranial disproportion in shunt-induced slit ventricle syndrome with cranial vault distraction osteogenesis. Pediatr Neurosurg 49:187–192. https://doi.org/10.1159/000358924
30. Sandler AL, Daniels LB, Staffenberg DA et al (2013) Successful treatment of post-shunt craniocerebral disproportion by coupling gradual external cranial vault distraction with continuous intracranial pressure monitoring. J Neurosurg Pediatr 11:653–657. https://doi.org/10.3171/2013.2.PEDS12404
31. Ryoo HG, Kim SK, Cheon JE, Lee JY, Wang KC, Phi JH (2014) Slit ventricle syndrome and early-onset secondary craniosynostosis in an infant. Am J Case Rep 15:246–253. https://doi.org/10.12659/AJCR.890590
32. Beuriat PA, Paulus C, Grassiot B, Szathmari A, Mottolese C (2015) Cranial vault expansion with the split bone flap technique in shunt-related craniosynostosis. Rev Stomatol Chir Maxillofac Chir Orale 116:239–244. https://doi.org/10.1016/j.revsto.2015.06.006
33. Bhadkamkar MA, Albright SB, Wolfswinkel EM, Bollo R, Buchanan EP (2015) Posterior cranial vault distraction in the treatment of shunt-induced craniosynostosis. J Craniofac Surg 26:e70–e72. https://doi.org/10.1097/SCS.0000000000001319
34. Mohan N, Albright AL (2015) Cranial expansion for shunt-induced craniostenosis case report. African J Neurol Sci 33:58–63
35. Kim SA, Letyagin GV, Danilin VE, Sysoeva AA Shunt-induced craniosynostosis: topicality of the problem, choice of the approach, and features of surgical treatment. Zh Vopr Neirokhir Im N N Burdenko 81:45–55. https://doi.org/10.17116/neiro201781445-55
36. Golinko MS, Atwood DN, Ocal E (2018) Surgical management of craniosynostosis in the setting of a ventricular shunt: a case series and treatment algorithm. Childs Nerv Syst 34:517–525. https://doi.org/10.1007/s00381-017-3648-y
37. Di Rocco C, Velardi F (2003) Acquired Chiari type I malformation managed by supratentorial cranial enlargement. Childs Nerv Syst 19:800–807. https://doi.org/10.1007/s00381-003-0837-7

38. Cornelissen MJ, de Goederen R, Doerga P, Cuperus I, van Veelen ML, Lequin M, Govaert P, Mathijssen IMJ, Dudink J, Tasker RC (2018) Pilot study of intracranial venous physiology in craniosynostosis. J Neurosurg Pediatr 21:626–631. https://doi.org/10.3171/2018.1.PEDS17480
39. Kraemer MR, Sandoval-Garcia C, Bragg T, Iskandar BJ (2017) Shunt-dependent hydrocephalus: management style among members of the American Society of Pediatric Neurosurgeons. J Neurosurg Pediatr 20:216–224. https://doi.org/10.3171/2017.2.PEDS16265
40. Sainz LV, Hockel K, Schuhmann MU (2018) Chronic overdrainage syndrome: pathophysiological insights based on ICP analysis: a case-based review. Childs Nerv Syst 34:401–408. https://doi.org/10.1007/s00381-017-3650-4
41. Barami K (2016) Cerebral venous overdrainage: an underrecognized complication of cerebrospinal fluid diversion. Neurosurg Focus 41:E9. https://doi.org/10.3171/2016.6.FOCUS16172
42. Freimann FB, Schulz M, Haberl H, Thomale U-W (2014) Feasibility of telemetric ICP-guided valve adjustments for complex shunt therapy. Childs Nerv Syst 30:689–697. https://doi.org/10.1007/s00381-013-2324-0
43. Beez T, O'Kane R, Piper I, et al. (2016) Telemetric intracranial pressure monitoring in syndromic craniosynostosis. J Craniofac Surg. https://doi.org/10.1097/SCS.0000000000002556
44. Martini M, Klausing A, Lüchters G, Heim N, Messing-Jünger M (2018) Head circumference - a useful single parameter for skull volume development in cranial growth analysis? Head Face Med 14:3. https://doi.org/10.1186/s13005-017-0159-8
45. Wright CM, Emond A (2015) Head growth and neurocognitive outcomes. Pediatrics 135:e1393–e1398. https://doi.org/10.1542/peds.2014-3172
46. Gault DT, Renier D, Marchac D, Jones BM (1992) Intracranial pressure and intracranial volume in children with craniosynostosis. Plast Reconstr Surg 90:377–381
47. Radic JAE, Vincer M, McNeely PD (2015) Outcomes of intraventricular hemorrhage and posthemorrhagic hydrocephalus in a population-based cohort of very preterm infants born to residents of Nova Scotia from 1993 to 2010. J Neurosurg Pediatr 15:580–588. https://doi.org/10.3171/2014.11.PEDS14364