Chapter

Will CSF Diversion in Patients with Idiopathic Intracranial Hypertension (IIH) Lead to Long-Lasting Shunt Dependency?

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Abstract

Long-term shunt dependency rates in patients treated for IIH with CSF diversion have not been established. We therefore present our experience with 5 children shunted for IIH during the years 1984–2000 with very long-time follow-up. Three out of these patients have experienced late or very late episodes of severe shunt failure during the second or third decade after initial shunt treatment. They were all boys and may not be representative for IIH patients as a whole. Three of them appear, however, to be permanently shunt dependent, indicating that long-term shunt-dependency in children treated for IIH with CSF diversion may be more common than previously expected.

Keywords: idiopathic intracranial hypertension (IIH), children, CSF diversion, shunt dependency

1. Introduction

Main topics

i.e. Shunt failure
Shunt revision
Shunt dependency/independency
Overdrainage
Shunt surviva

In 1937 Walter Dandy published a series of 22 patients treated surgically for intracranial hyper-tension without a tumor [1]. Among his cases treated with subtemporal decompression during a period of 10 years, there were 3 children. Later on, this condition with papilledema, headache and visual disturbances was called benign intracranial hypertension, and many of them could be managed favorably by medical treatment [2]. In more severe cases, it soon became clear that this obscure disease is not always that benign [3]. In severe cases the term malignant pseudotumor was introduced [4]. Later on the term Idiopathic Intracranial Hypertension (IIH) was introduced [5].

Neurosurgical treatment with CSF diversion has been performed in severe cases where the response to medical treatment has been unsatisfactory, or as primary treatment in patients with severe visual affection [6, 7]. Unlike the situation for

hydrocephalic children treated with CSF shunts who for the most become shunt dependent, clinical results on long term shunt dependency in IIH are unavailable.

Modified Dandy criteria:

Symptoms and signs of raised intracranial pressure (headache or papilledema).
No localizing signs in neurological examination (except abducent nerve palsy).
Normal neuroimaging.
Increased intracranial pressure as measured by lumbar puncture.
Normal CSF composition.
Alert and awake patient.
No other cause of raised intracranial pressure.

1.1 Diagnostic work-up

Idiopathic intracranial hypertension is a diagnosis where other causes I leading to increased intracranial pressure have been excluded by CT or MRI, including unobstructed venous outflow. Normal CSF composition rules out infection. The diagnosic procedure of intracranial hypertension itself is usually performed by a simple lumbar puncture. This gives CSF for analysis, and fluid may spurt out in cases of severe IIH. When the Intracranial pressure (ICP) is measured via the lumbar route, the patient is posisioned flat with the puncture site in equal hight as the IIIrd ventricle. The needle seize should not be very thin (G19), to obtain a presice manometric pressure reading. Loss of fluid should be avoided, since the pressure falls rapidly unless ICP is very high. Pathological elevated ICP is above 25 cm H_2O (15–20 in children). ICP is,however, influenced by a number of factors that may give misleading results when ICP is evaluated by the lumbar route.

Firstly, the patient should be cooperative and relaxed. Furthermore ICP is highly influenced by changes in $PaCO_2$. If the patient is anxious and hyperventilate, ICP will fall rapidly. The pressure recording can also be performed by the use of standard fluid pressure transduces and may be combined by other diagnostic work-up, such as lumbar infusion tests. In children, these diagnostic procedures must be undertaken during general anesthesia and normocapnia.

When patients with severe headache but without papilledema or visual affection, are given a IIH diagnosis, correct evaluation of the CSF hydrodynamic situation is pertinent. If CSF pressure is observed under unstable conditions or is influenced rapid changes in the central venous pressure, misinterpretations may take place. Queckensteds test (venous compression in the neck) demonstrates this effect on the CSF pressure, and was previously used to exclude a spinal block. Lumbar infusion tests can be used to explore the need for therapeutic CSF diversion, and of course demonstrate that an indwelling shunt is patent (LP-shunt or VP-shunt). A simple lumbar puncture will of course also be helpful to exclude severe shunt failure, as well as shunt infection.

It should be pointed out that the lumbar puncture itself will give an opportunity to explore the possibilities for a good result in IHH patients. After puncture with a 19G cannula, CSF will leak during the following three weeks, especially in true IIH patients with increased CSF pressure. This is why some individuals without IIH, will experience post-puncture headache due to intracranial hypotension. The beneficial effect of LP and CSF removal on severe headache and visual affection, is often temporary and repeated punctures or shunt implantation will be needed.

2. Methods

All children (0–19 years old) who underwent a shunt implantation for IIH during the years 1980–2000 in our institution were identified from the surgical

protocols. Ethical approval was obtained from the medical ethics committee of Norway, the Regional Committee of Medicine and Heath.

We present our experience with these pediatric patients shunted for IIH during the years 1984 to 2000; and therefore have long-term follow-up.

3. Results

Five pediatric patients aged 0–19 years presented with clinical signs and symptoms of increased ICP.

They had small ventricles, no venous outflow obstruction, high opening pressure on LP and normal CSF composition.

They were all males, four were in the first decade (age: 1,3,4,5 and 18 years). All had bilateral papilledema, headache and visual disturbances in spite of medical treatment (Acetazolamide, corticiosteroids, furosemide). They all had normal weight for age. The results are summarized in **Table 1**.

Patient number Age/sex	Year	Shunt procedure	Number of shunt proce-dures	Follow-up years	Shunt depend?	Clinical presentation
1 M 18	1984	LP	3	36	Yes	Headache, visual loss, papilledema
	2003	LP(rev)				Headache, blurred vision
	2013	VP				Headache
2 M 4	1988	CA		32	No	Papilledema, VIth n. palsy, visual loss
	2016	Remove				Overdrainage symptoms
3 M 3	1989	CP	3	31	Yes	Papilledema, visual loss
	2006	LP				Headache, vomiting, High CSF pessure
	2009	LP(rev)				Headache
4 M 5	1991	CP	5	29	Yes	Hedache, papilledema, ataxia, blurred vision, unstable level of consciousness
	1994	revCP				Headche, vomiting
	2004	VP				Headache, vomit, diplopia,papilledema
	2006	LP				Episodes of vislual loss, high ICP
	2009	Remove P-kat				Abdominal disability
5 M 1	2000	LP	1	18	?	Blilat. VIth nerve palsy, papilledema

LP – lumboperitoneal; VP – ventriculoperitoneal; CA – cisterno(Magna)atrial; CP - cisternoperitoneal.

Table 1.
Clinical details.

3.1 Case 1

The first patient was a 18 year-old boy. In spite of medical treatment and LP every 4 weeks, the headache was troublesome and his vision was partly lost (0,5)

during six months in spite of the repeated taps. A lumboperitoneal shunt (mini Holter high valve) was therefore implanted in January 1984. The symptoms resolved and his vision improved but not completely during the next year. After many years, in 2003, he once more experienced headache and blurred vision. There was no papilledema, but high opening pressure on LP. The LPshunt was found blocked and after revision he once more was symptom free.

In 2013 another episode of clinical shunt failure took place, and the symptoms again responded favorably after implantation of a VP shunt. Today, with 36 years follow-up, he is in full-time work as a diary worker.

3.2 Case 2

The second patient was a 4 year-old boy presenting with VIth nerve palsy, bilateral papilledema, headache and visual affection in spite of treatment with furosemide for some weeks. After implantation of a cisterno-atrial shunt (mini-Holter high valve), the symptoms resolved (slight binasal visual defect). After many good clinical years until about 2012, he thereafter during the next years experienced increasing overdrainage-symptoms (headache, dizziness, sometimes also subjective affection of vision and hearing) when rapidly changing to the upright position. The symptoms were reversed effectively in the recumbent position. In 2016 the shunt was removed and ICP was monitored in 4 days along with clinical observation. He clearly improved, and his clinical symptoms related to change in body position disappeared. He now appears to be shunt independent for more than three years.

3.3 Case 3

The third patient was a 3 year-old boy admitted with a short history of lost vision for 36 hours, reduced pupil reactivity, ataxia and poor general condition. Fundoscopy demonstrated choked discs and cerebral MRI was normal including unobstructed venous outflow. Lumbar puncture revealed normal CSF composition and increased CSF pressure, but the ICP level was difficult to measure due to lack of cooperation. A lumbar infusion test during general anesthesia demonstrated increased CSF opening pressure as well as slightly increased outflow resistance.

Due to the dramatic clinical symptoms with complete visual loss, corticosteroid treatment was implemented and an acute shunt procedure was performed during the same general anesthesia. A proximal catheter was introduced into cisterna Magna and connected to a low pressure Holter valve with diversion to the peritoneal cavity.

His vision gradually reappeared within days, and after one week there was normal vision and pupillary reactivity to light. Some ataxia and clumsy motor function persisted for weeks, but after 6 months his clinical condition was quite normal. During the first two years of treatment he experienced a few episodes with headache, ataxia and diplopia (VIth nerve paresis) which resolved spontaneously within a couple of days or after pumping on the Holter valve. At the age of 5 years (1991) he demonstrated episodes of overdrainage in the upright position, which subsided after implementation of an ASD (anti-syphon-device) distal to the valve.

Thereafter his clinical condition was uneventful for many years. During his university studies (in 2006), he became acutely ill with signs of increased ICP (headache and vomiting). There was no choked discs, but lumbar puncture revealed markedly increase CSF pressure level (50 cm H_2O) and no signs of infection. Once more MRI was normal (**Figure 1**). After a shunt revision (LP-shunt), his clinical condition normalized within a few days. In 2009, he experienced shunt failure once more, again followed by rapidly improvement after shunt revision.

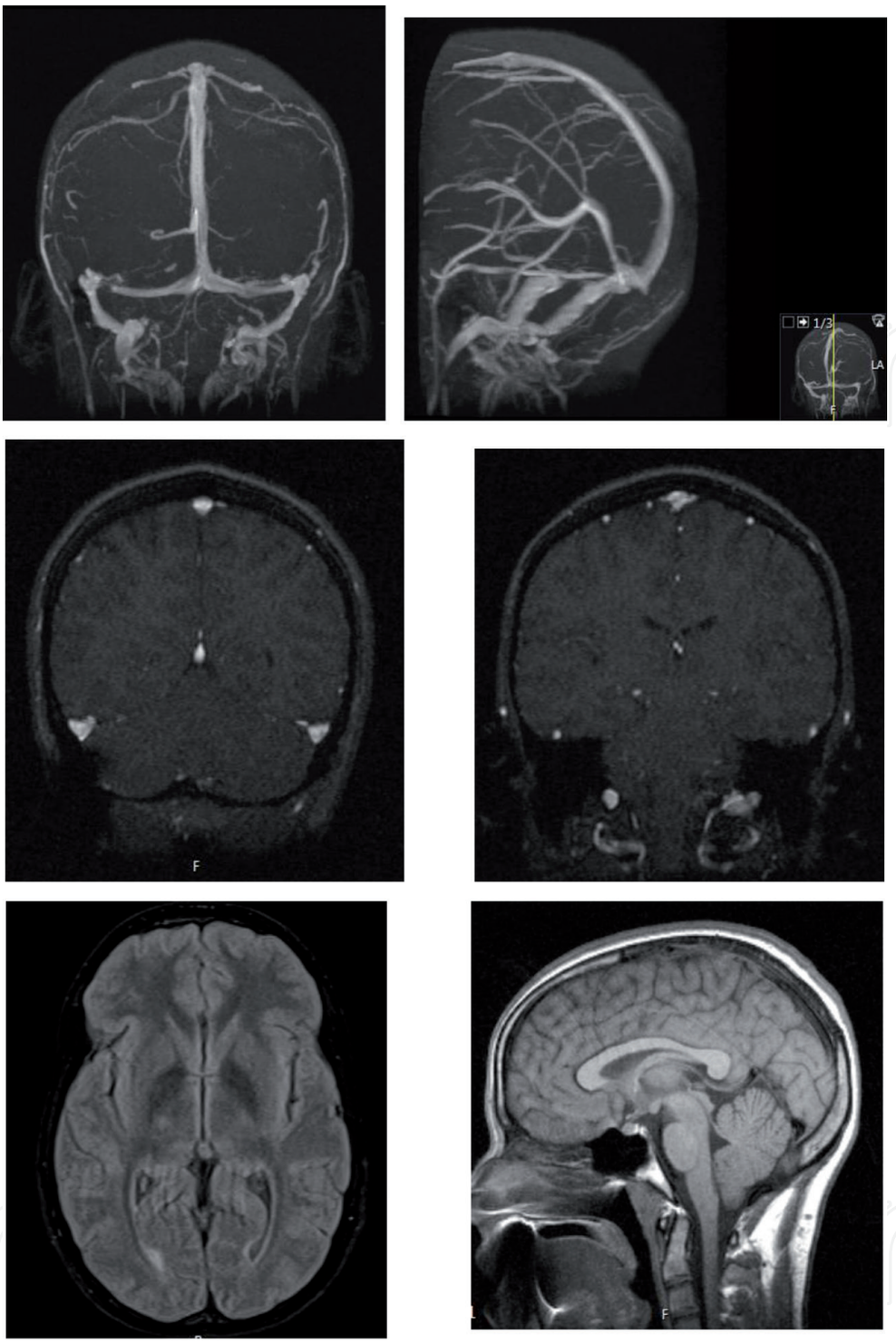

Figure 1.
(Case 3) demonstrates normal intracranial structures during the episode of shunt failure in 2006 (lower row) including unobstructed venous outflow (upper and middle row).

Thereafter he has been working full time, and has been clinically quite well for another 11 years.

This case with a very rewarding clinical result indicate that this 35 year-old man in excellent condition most likely is permanently shunt dependent.

3.4 Case 4

The fourth patient was a 4 year-old boy presented with headache, bilateral papilledema, ataxia, and unstable level of consciousness and episodes of blurred

vision. The ventricles were small, venous outflow was unaffected and a lumbar infusion test during general anesthesia (normocapnia) revealed ICP of 24 mm Hg and increased resistance to outflow of CSF. A medium Holter shunt was implanted from cisterna Magna to the peritoneal cavity and the symptoms normalized. Three years later he experienced acute shunt failure(disconnection) with headache, papilledema, vomiting and a lumbar pressure of 45 mm Hg. After revision, the situation once more normalized. Ten years later, in 2004, he experienced another episode of shunt failure with headache, vomiting, diplopia and bilateral papilledema. Lumbar pressure was highly increased. A VPshunt (Codman Medos) was now implanted. During the next two years the situation was more unstable, with episodes of clinical symptoms related to over- as well as underdrainage. In 2006, he experienced episodes of visual loss, and ICP monitoring revealed pathological ICP. A lumboperitoneal shunt (OSV II) was therefore implanted and the VPshunt removed. During the subsequent 12 years he has been clinically well and fulfilled a master degree at the university.

3.5 Case 5

The fifth patient was a one-year-old boy with bilateral papilledema, bilateral VIth nerve paresis and a LPshunt was implanted in 2000 with good clinical response. His development during the following years has been uneventful. He is now working as an electrician, climbing in poles and without any clinical symptoms of ICP disturbances.

4. Discussion

Clinical series of patient with IIH are heterogenous with respect to age, sex and the severity of disease. When Dandy presented his series of 22 cases in 1937, he gave an individual and detailed overview of clinical history, symptoms and signs of his patients [1]. They illustrate the severity of the problems these patients were faced with when they sought neurosurgical treatment 80–90 years ago. One of 3 children in Dandys` series was:

4.1 Dandy's case 12

Female Age 13 –January 21 to February 1936.
Complaints. – Failing vision, diplopia and headache.
Present illness – Seven months ago pain developed in her left hip causing her to limp. The pain progressed steadily for three months when she was no longer able to walk. Roentgenograms were taken and were said to have been negative. A month later a second roentgenologic examination revealed an abnormality about the epiphysis of the femur and atrophy of its neck. The leg was placed in a plaster spica. The pain immediately disappeared. She felt better, ate heartily and gained some weight. A month later, i.e., two months ago, she had an attack of vomiting. One month later, i.e., one month ago, she complained of dizziness and headache over both eyes. Within a week her eyesight became blurred, there was double vision and the headache had become much more severe. It was then located in the occipital as well as the frontal region. There were pain and stiffness in the neck. Vomiting became more severe, occurring several times a day. Three weeks ago the plaster spica was removed and an appendicectomy performed because of the vomiting. There was no upset following the operation, and although her headache continued, the vomiting ceased. One week ago her vision had become so poor that she could only recognize light with the left eye. She was still able to read with the

right eye. For the past three or four weeks there have been attacks of numbness in the right (not the leg in the spica).

Examination.- Patient is a sallow, fairly well nourished, young girl suffering severely with headache. Temperature normal; pulse 110; respirations 24; blood pressure 120; W.B.C. 7,800. There is a definite cracked-pot sound (Maceven's sign) on tapping the frontoparietal suture line. Moreover, roentgenologic examination showed separation of the sutures – unusual at the age of 13, and indicative of an extreme degree of intracranial pressure. There is only light perception in the left eye. She can read ordinary print with the right eye. Being bedfast and in a plaster spica, a more detailed eye examination is not possible. There is papilledema of five to six diopters in the right eye; two to three diopters in the left (the blind eye). The disk and surrounding retina are filled with large flame-like hemorrhages; these are more pronounced on the right side. There is weakness of the external rectus muscle on the left, but the parents say this has always been present. The knee jerks on the right could not be elicited; The left leg is in a plaster spica. Babinski is negative; no clonus.

Diagnosis. – Although I had suspected a tuberculous hip and a metastatic infection of the brain, Dr. George Bennett, who saw her with me, excluded tuberculosis from the study of the roentgenograms. The coexistence of the two lesions made us suspect a relationship between the two, but the only positive finding in the hip was the epiphyseal separation and atrophy of the neck and upper part of the shaft. There was no positive infective process.

Trephine and Air Injection. – January 22, 1936. The right ventricle was tapped. Fluid spurted out under tremendous pressure – at least a distance of three feet. About 15 cc. of fluid escaped and then the flow shut down abruptly. Ten cubic centimeters of air were injected under pressure to replace the fluid. The ventriculograms showed a perfectly normal ventricular system. The fluid showed four cells, all lymphocytes. A guinea-pig was inoculated with the fluid, because of the suspicion of tuberculosis; it had no effect upon the animal.

A right subtemporal decompression was performed immediately after the ventriculograms had been interpreted. The dura was exceedingly tense. A small nick was made in the dura, hoping that fluid might be encountered and thus reduce the terrific tension. A large amount of fluid did escape, but it seemed to make little, if any, impression upon the tension of the dura. The dura was rapidly opened but the pressure was still so extreme that the cortex ruptured inferiorly. The intracranial pressure has just about reached its limit.

Following the operation the decompression was exceedingly tense. A spinal puncture on the third day after operation registered 460 Mm of water; this, in spite of decompression. With this great pressure it looked as though the decompression would be futile. A lumbar puncture was performed on each of the following seven days; about 30 cc. of fluid being removed each time. On the eighth day after operation the spinal fluid pressure registered 350 Mm. The tension of the decompression gradually decreased during the next five days. On the fourteenth day the decompression was flat and the spinal fluid pressure measured 160 Mm. Patient remained in the hospital a week longer. The decompression remained perfectly flat throughout that time.

For a few days after operation patient was unable to se with either eye. As the pressure became less her vision returned and at the time of her discharge she was able to read fine print with the right eye, but the left eye still remained blind. Her general condition had changed entirely, her color was better, and she was very much more alert and active mentally.

Subsequent Course. – When examined by me three months later, she was totally blind, had severe headaches, and the decompression was full and as tense as it could possibly be. The left optic disk showed extreme optic atrophy with sharply defined disk and normal sized veins. The right had much the same appearance but slightly less advanced. It did not look as though vision could ever return. Within two weeks the decompression was again flat, and vision returned on the right eye,

February 12, 1937. 13 months after the operation, she was well; had no more evidence of increased pressure, the decompression had remained soft; her vision was 20/70

in the right eye and there was a fairly normal field of vision. Her femur has healed and gives no trouble; there is no limp.

4.1.1. Historical perspective on IIH management

Patients with extreme presentations in terms of visual disturbancies and headache underwent subtemporal decompression procedures in small volumes during the following decades. Although positive clinical responses were obtained in shortly after decompression in many of them, the procedures were dangerous and serial lumbar punctures had to be performed to achieve more long lasting relief from the decompression. When lumbo-peritoneal shunting was introduced in the treatment of hydrocephalus in the 1950ies, it was therefor natural to explore this treatment also in severe cases of pseudotumor. When other CSF diversion procedures for HC were established in the 1960ies (VA-shunts) [8], and later on VP-shunts [9], they were also found applicable in treatment of iIH.

Although shunt procedures are generally effective in relieving the CSF pressure in IIH, these CSF procedures have a substantial number of complications. When medical treatment for IIH became available (Diamox, Furosemide etc), non-surgical treatment would be preferred by many, since shunt failure, shunt infection and shunt dependency could therefore be omitted.

During the 1980ies and 1990ies, the number of patients with IIH increased rapidly, especially among people with severely increased body mass index, with a clear female dominance [10].

In some of these patients bariatric surgery has been introduced to reduce the weight problem and may also solve the IIH or alt least reduce the need for CSF diversion procedures [11]. It also seems that morbid overweight increases the problems with CSF diversion to the peritoneal cavity.

Several studies have compared lumboperitoneal shunts versus ventriculoperitoneal shunts in the treatment of IIH [12]. Both types of CSF are highly effective in the acute stage, but there are more revisions during the following years in patients treated with LP-shunts [13]. McGirt and coworkers furthermore underscore that while CSF diversion is effective in the acute stage, several patients experience that headaches returns. The found that this happens more often in patients without papilledema and long-standing symptoms before shunt treatment.

In early years LP shunts were considered applicable since it was technically easy to achieve CSF from the lumbar CSF reservoir, compared to get ventricular access in patients with small or slit ventricles.

In recent decades modern imaging (MR,CT) and stereotactic placement of ventricular catheters have resulted in ventriculo-peritoneal CSF shunting as the most used procedure, at least in adults. In patients with severe overweight, distal end problems with abdominal complaints is common.

Pediatric IIH is a rare disease [14] and pediatric cases are often presented in series of institutional IIH patients of all ages, where females of reproductive age dominate. Another problem is that clinically IIH covers a wide spectrum of symptoms and signs. When patients without papilledema or frank visual affection are included, non-surgical management is preferred and may lead to satisfactory outcomes. Long-term follow-up of IIH-children treated conservatively (with papilledema) has demonstrated that some are left with visual defects [15]. In recent years focus has been directed to the small group of IIH patients where troublesome headaches are not the main threat, but visual affection. If treatment over time is not efficient in reducing papilledema and visual affection, permanent reduction in visual acuity, visual field defects and even blindness may take place [16, 17].

This was the case in our Case 1, where serial lumbar taps gave excellent symptomatic relief of headache for 3 weeks, but during the fourth week the symptoms built up. When a LP-shunt eventually was implanted after 6 months, his visual acuity was reduced by 50%, and only improved slightly during the subsequent year. He obviously should have been shunted earlier. The visual threat also have a strong time component, implying that when visual deterioration is rapid or abrupt, surgical intervention is imminent (CSF-diversion or optical nerve sheath fenestration). Progression of visual affection over weeks have been considered a strong indication for surgery, with the prospect of avoiding further visual affection. Our Case 3, demonstrates that frank blindness can be treated successfully with restoration of vision, but this raises the question of emergency surgery in such rare cases. A number of publications point to this problem when patients with IIH are faced in emergency departments [6, 18,–20]. These rare, but very important experiences are in accord with the eminent and detailed clinical observations made by Walter Dandy 80 years ago.

Over-night blindness has even been reported after shunt failure in IIH [21]. Shunt failure may result in reoccurrence of sever headache and visual affection, and even lead to CSF leak [22].

Thus, neurosurgical treatment by CSF diversion is well established in IIH patients with severe visual affection as well as in cases with unsatisfactory response to medical treatment [23]. When the clinical result is good over years, as in our patient 5, it is difficult to know if the shunt is still functioning or the underlying condition has normalized spontaneously [24].

However, our five cases with very long-term follow up (20–36 years) demonstrate episodes of shunt failure in three of them, up to 29 years after initial shunt treatment. All together 7 episodes of shunt failure requiring shunt revision were observed. Only one of these distinct shunt failures took place during the first decade after implantation (after 3 years, disconnection). The others occurred from 13 to 29 years after implantation. All the shunt failures were restricted to 3 out of the 5 patients, and all of these 3 experienced repeated delayed shunt failures (in the second or third decade after initial treatment). There are few pediatric series with shunt treated IIH children reported. Niotakis and coworker report on 7 cases who underwent lumbar CSF shunt surgery. There was 2 acute procedures due to severe visual affection, but 3 patients presented with severe headache without papilledema. All patients had shunt revisions, due to symptoms of overdrainage or obstruction. An adult series from Dublin [25] reported low failure rates both in LP-shunts (11%) and VP shunts (14%), nevertheless revision rates were 60% in LP shunts and 30% in VP shunts. Bjørnson and coworkers report a lower revision rate (11%) in VP-shunted IIH adults. The follow-up periods in these studies are restricted, and late shunt failures have not been reported in adults (beyond 7 years).

Since IIH has been considered a benign and perhaps self restoring condition, this might explain the lack of long-term follow-up studies. Good long-term results can therefore both be explained by a no-longer existing CSF diversion demand andsome of the well functioning shunted patients may in fact have functioning shunt. This question has not been addressed, but very late shunt failure has not been reported. Many revisions in reported in IIH shunted individuals, describe other problems than shunt failure (obstruction or hypofunction); i.e. overdrainage, shunt infection or unstable diversion.

Shunt implantation in children with severe IIH raises the question of making these children permanently shunt dependent [3]. While some authors have addressed this question, no cases with late or very late shunt failure has been reported, until our case 3, published in 2017 [26]. Four of our pediatric cases have

been published previously [27], and our case 1 was not included since his age of 18 did not fit with the series (reviewers opinion).

This case with 30 years follow-up, indicate that individuals shunted for IIH can be persistently dependent on their shunt, and may experience acute shunt failure even after many years of treatment. We also consider our patient 1, with shunt failures 19 and 29 years after initial treatment to be permanently shunt dependent. Also patient 4 with shunt failure after 15 years is most likely in the need for life-long shunt treatment.

The last two patients tell a different story. Patient 2 had a good clinical response to shunt treatment in 24 years and thereafter experienced progressive symptoms from overdrainage. His shunt was therefore removed under ICP observation for 4 days, and he responded well. After 3 years further follow-up, we consider him shunt independent.

Our small group of shunt treated IIH children is peculiar since they are all boys, compared to the well known female dominance in IIH patients as a whole. It is, however, also known that females do not outnumber males in the first decade in IIH treated children [28]. In a report from a large pediatric emergency unit, children with papilledema and visual affection diagnosed with IIH, were twice as common in boys compared with girls in the first decade of life, while the opposite was found in the second decade [19]. It is unclear if our small study group is representative for all pediatric IIH patients and for the large group of adult IIH patients, many with high body mass index (BMI).

Clinical studies describing adult IIH cohorts illustrate the female dominance, often up to 90 per cent.

Comparison of treatment strategies as well as results is troublesome because inclusion criteria may vary. In the United States a rapidly rising incidence of cerebrospinal fluid shunting procedures for idiopathic intracranial hypertension was described for the years 1988–2002 [10]. The increase seemed to correlate with the rapid rise in morbid obesity. The increase was 320% for the population as a whole, but only 38% for males, and 52% for pediatric cases (<13 yrs., both sexes).

Bariatric surgery is considered to be beneficial for overweight IIH patients, and may reduce the need for shunt treatment. Bariatric surgery has been used as primary treatment as well as additional surgical procedure in IIH patients after CSF shunt procedures. The are a number of reports on multiple shunt procedures in such patients. In a group of 6 patients where bariatric surgery was performed in shunted IIH individuals, the shunt was externalized and clamped. Due to severe overdrainage symptoms, shunt removal was planned, but the shunt had to be acutely reestablished due to unexpected and clear shunt dependency [29].

This points to another problem, namely that evaluation of shunt function in IIH patients can be difficult. When headache is the dominating clinical problem, differentiation between high-pressure symptoms (shunt failure), visual affection; and low-pressure symptoms (over drainage) may be challenging. It is easy when overdrainage-symptoms in terms of headache and dizziness are rapidly reversed in recumbent position, but if the CSF drainage is unstable the clinical picture may include both. In such situations ICP-monitoring or use of lumbar or ventricular infusion tests may be helpful [27, 30], and additional shunt revisions and increased risk of shunt infection can be reduced.

Although the report from Roth and coworkers [20] presented 6 aldults that were unexpectedly shunt dependent, it is remarkable that long-term shunt dependency has not been a topic. Since benign intracranial hypertension has been considered a self-solving condition, long lasting shunt dependency may therefore have been unexpected, but perhaps also overlooked. In research on shunt patency in hydrocephalic patients, the term *shunt survival* has been used; meaning the time from the

initial shunt procedure to the first shunt revision. This has been a kind of quality measurement in management of hydrocephalic patients, and presented in per cent for the first, second or five years.

The proportion of patients still having their initial shunt unrevised was anticipated to represent individuals with persistent shunt dependency, but harboring still functioning shunts.

In our long-term follow-up of children shunted for hydrocephalus I the 1960is and 1980 we have found shunt dependency rates of about 90% [2, 11]. That is why ETV is now a preferred treatment when feasible in primary management of hydrocephalus. The other 10% are for sure not shunt dependent, since their shunt has been explanted, in some of them in combination with an ETV procedure. The correct description would therefore be that up to 90% still are or can be more or less shunt dependent.

5. Conclusion

The shunt dependency rates for pediatric IIH treated with CSF diversion are actually not known, and long-term follow-up reports are scarce. Our limited experience, however, indicate that permanent shunt dependency may be more common than previously expected. In adults this topic has hardly been addressed.

Conflict of interest

None.

Abbreviations

CSF	cerebrospinal fluid
LP	lumbar puncture
IIH	idiopathic intracranial hypertension

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