Acta Neurochir (Wien) (1992): 116: 155–160

Acta Neurochirurgica

Surgery of Hydrocephalus: Past, Present and Future

J. F. Hirsch

Service de Neurochirurgie Pédiatrique, Hôpital Necker Enfants Malades, Paris, France

Summary

The treatment of hydrocephalus, over the centuries, underwent three stages of evolution.

During antiquity, middle ages and Renaissance, hydrocephalus was not understood. Medical treatment was useless; surgery was hopeless.

The second stage extends from the XIXth century to the end of the first half of the XXth century. CSF circulation was now understood; surgery however, remained inefficient, but some patients survived with arrested hydrocephalus.

The third stage begins in the nineteen fifties with the development of silicone shunts with a valve. Surgery transforms the prognosis of hydrocephalus, but the number of post-operative complications creates new problems. The different attempts that have been made during these past two decades to solve these problems are reviewed. They have resulted in a reduction of the mechanical and infectious complications. CSF overdrainage has been minimized. Percutaneous ventriculocisternostomies have in some cases replaced shunts.

In the future, to improve outcome in these hydrocephalics, surgery, when indicated, should be performed as early as possible. Knowledge and prevention of the causes of hydrocephalus should be developed.

Keywords: Hydrocephalus; ventriculo-cisternostomy; shunt complications; CSF overdrainage.

Any progress in Medicine implies the answer to two questions: where are we coming from? What is the present stituation? The evaluation of the present shows what is well and what could and should be improved. Then questions being clear, solutions can be looked for.

The treatment of hydrocephalus, over the centuries, underwent three stages. The first stage before the knowledge of the CSF ciculation, covers the period from antiquity to the 18th century. The second stage extends from the XIXth century to the end of the first half of the XXth century. The third begins with the second half of the XXth century and is characterized by the development of CSF deviation system with valves.

I) During antiquity and the middle ages, hydrocephalus was known, but not yet understood. Hippocrates knew that the accumulation of water within the head caused it to enlarge. Oribasius and Paul of Aegina thought that the fluid was located between the scalp and the pericranium, the pericranium and the bone or between the skull and the dura. Vesalius in the 16th century was the first to show that the water was accumulating within the ventricles. Ambroise Paré stated that, when the water was within the ventricles, recovery was not possible. Surgery of hydrocephalus was, in these times hopeless. However several authors such as Charaf ed Din (Fig. 1), William of Salicet had tried surgically to remove water from the ventricles. The results were catastrophic. L. Heister states: "If you

Fig. 1. Charaf Ed Din (1465). Removing surgically, "humidity" from the head of a child

* Invited Lecture, presented at the European Congress of Neurosurgery, Moscow, June 23–29, 1991.

make a paracentesis on the head to discharge the lymph, your operation is no sooner performed, but the infant dies, as physicians have been too often assured by experience". Herrissant in 1772, in his "Dictionnaire Universal et Raisonné de Médecine, de Chirurgie et de l'Art Vétérinaire", writes: "Il serait aisé, dans quelques cas, de pratiquer la ponction ou des scarifications; mais les épreuves qu' on en a faites, n'ont pas été heureuses". The treatment is therefore purely medical: diuretics to remove the fluid "per urinam", sponges placed on the head, bandages, diaphoretics such as salt of deer horn or viper's powder. It is even stated in Herrissant's dictionnary that, if the patient wants to drink some wine, he may do so. The age of the patient is not specified.

II) The second stage in the treatment of hydrocephalus begins at the end of the 18th century and in the beginning if the 19th with the progressive understanding of the CSF circulation. Cotugno describes the cerebrospinal fluid, Monro and Magendie the CSF pathways. The most important publication is that of Dandy and Blackfan[2] who, in 1914, give an accurate description of the CSF circulation and of its pathology. This progress stimulates the development of surgery, essentially during the first half of the XXth century. Most operations however usually end up in failure. Intracranial bypasses through an open craniotomy, such as the perforation of the lamina terminalis, often block rapidly; the post-operative mortality is not negligible in the Torkildsen's operation[37]. The proper material is not available for extracranial bypasses. The consequences of Matson's operation[22] in which, after a nephrectomy, the CSF is diverted through the ureter towards the bladder, are too great.

The results of the treatment of hydrocephalus during that first half of the XXth century can be evaluated

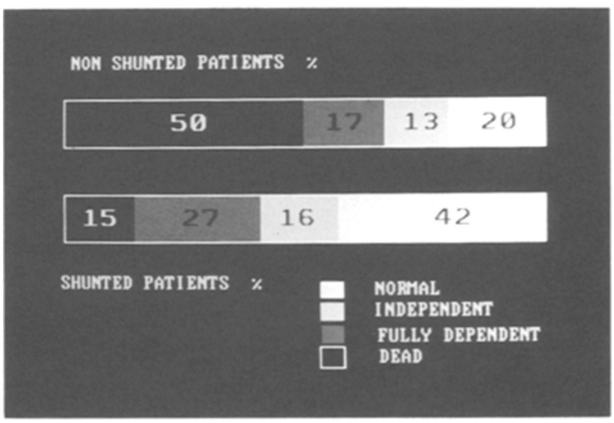

Fig. 2. Overall mortality and functional results. Before the shunt era: upper line; with shunts: lower line

through the puplications of Yashon[37], Hagberg[8], Laurence *et al.*[17, 18] and Jansen *et al.*[13, 14]. The overall mortality is about the same in these different series, varying from 45% to 53%. In other words, half of the patients die. Actually death in hydrocephalus is a time related event so that the curve of the survival rate given by Laurence allows a better evaluation of the real situation. It shows that 10 years after birth only 20% of the patients are still alive. For the assessment of outcome, patients are separated into three groups in those four series: able to work, independent and dependent. The results are about the same in the different series. The mean values are 43% able to work, 25% independent and 32% dependent. These results are however biassed by the fact that they take into account the patients who survive without any treatment and who probably present a type of hydrocephalus of slow evolution or an arrested hydrocephalus.

A rough summary of the results in that period might be the following. Out of 100 hydrocephalic children, 50 die, 20 are able to work, 13 are independent but can not work and 17 are dependent (Fig. 2).

III) The second half of the XXth century is a turning point in the treatment of hydrocephalus. After the work of Nulsen and Spitz[25] and of Pudenz *et al.*[28], after the introduction of the Holter valve in 1956, the treatment of hydrocephalus becomes really efficient. Silicone shunts with a valve are inserted in most patients.

The overall mortality decreases abruptly to about 15% (Table 1).

The actuarial survival rate, in the series of "Les Enfants Malades", remains stable after the first years. The improvement is therefore obvious.

Outcome has also improved, but is far from normal. 52% of the IQs in our series are above 80, but 21% are between 60 to 80 and 28% are under 60. Severe behavioural disorders are observed in half of the children. Finally only 46% of the patients attend a normal school (Fig. 3).

Table 1

	% Mortality	Nb. of Patients
Amacher and Wellington[1]	21%	170
Lorber and Zachary[19]	17%	30
Mazza *et al.*[23]	18%	165
Keucher and Mealey[15]	16%	228
Hirsch *et al.*[10]	11%	576
Total	17%	1169

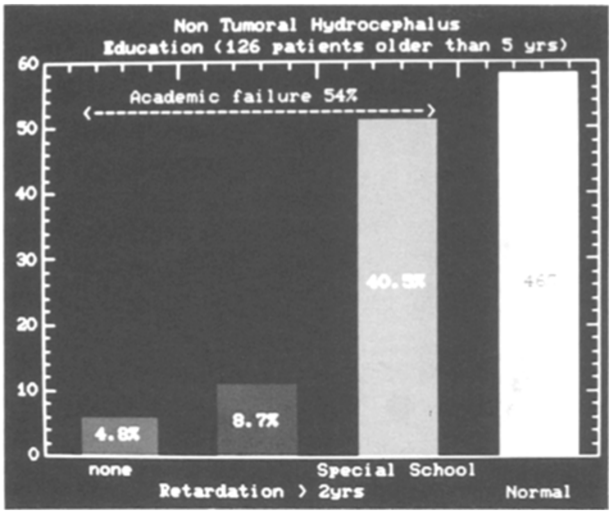

Fig. 3. Academic performance in hydrocephalics. Only half of the patients attend a normal school

To summarize these results and compare them with those of the pre-shunt period, out of hundred patients, 15 die, 42 are normal and able to work, 16 are independent but do not work and 27 are fully dependent (Fig. 2). The improvement is characterised by the diminution of the overall mortality and the increased number of normal children. However it should be pointed out that there is also an augmentation of the number of fully dependent patients, probably due to the fact that, in the pre-shunt era, these children would have died.

Another important criticism of the shunt technique is the number of post-operative complications. The rates of post-operative infection vary with the different authors, ranging usually from 5% to 10%. The rate of shunt failures due to other causes is important and time related. In our series ten years after the initial shunt insertion, more than 70% of the patients have been operated on at least twice for such a shunt failure[33].

This review of the results obtained with shunts in the treatment of hydrocephalus demonstrates clearly that not all problems are solved. The number of post-operative complications should be reduced; the outcome should be improved. We have tried to achieve this by re-establishing the normal CSF pathways without shunting everytime it was possible, by lowering the infection rate, by improving the design of the shunts, by developing more physiological new shunt systems.

IV) In some patients, it is possible to re-establish the CSF circulation without any shunt by removing an obstruction due to a cyst or a tumour or CSF hypersecretion caused by a papilloma. In aqueductal stenosis, some authors like Lapras[16] insert a simple tube between the IIIrd and the IVth ventricle. The solution that we have adopted in non-communicating hydrocephalus is the percutaneous ventriculocisternostomy. This technique was not new since it had been initiated by Mixter[24] in 1923 and then used by Scarff[34] and McNickle[21]. However this technique was really revived by G. Guiot[6] in 1963 under endoscopic guidance and in 1968[7] under X-ray guidance. G. Guiot used it in cases of late hydrocephalus due to aqueductal stenosis. We began to use this technique in 1973[26] and showed that it could also be used in infants with about 70% of good results when the patients were correctly selected. Jaksche and Loew recently demonstrated even 80% good results. The operation was at that time performed under X-ray guidance. We have recently changed the technique: all our operations are now performed under endoscopic guidance. This technique allows a perfect visualization of the foramen of Monro, of the mamillary bodies and of the floor of the IIIrd ventricle, in front of them, where the perforation is made (Fig. 4). So far, under endoscopic guidance, the post-operative mortality has been nil. The rate of post-operative infection is also practically nil. Secondary blockage of the ventriculostomy occurs in 16% of the patients, but the operation can be repeated. More importantly since normal CSF circulation is re-established, one can expect the best possible outcome, at least the outcome depending on the cerebral lesions

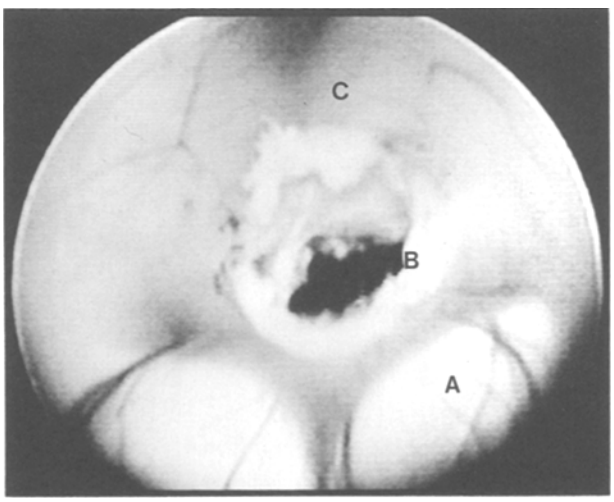

Fig. 4. Ventriculocisternostomy under endoscopic guidance. *A* Mamillary body; *B* Perforation; *C* Floor of the IIIrd ventricle. The perforation is performed in front of the two mamillary bodies, at the midline of the floor of the IIIrd ventricle

present before surgery, on the aetiology and the ventricular dilatation.

V) However shunts remain in most cases the unique solution for treating hydrocephalus, especially in communicating hydrocephalus, since the different attempts that have been made to cure the disease by cauterization of the choroid plexuses[3, 4, 29, 31, 35, 36] have been unsuccessful. Therefore to improve the present state means to improve shunts and shunt insertions in order to reduce the rate of post-operative infection and the rate of mechanical complications. Shunts can be improved in two different ways, in their design and in their function.

A) In order to reduce the rate of post-operative infection, we have tried to eliminate intra-operative contamination; we have studied the factors that favour infection, before or after surgery and we are now trying to determine whether prophylactic antibiotic therapy is useful or not.

To decrease intra-operative contamination, we have applied to shunt insertions a technique which was used in laboratories to breed germ-free animals[9]. The use of a surgical isolator reduced to nearly naught intra-operative contamination and divided by three our rate of acute post-operative infections.

Reviewing 1174 shunt insertions performed in "Les Enfants Malades"[30], we found that the risk of infection was larger in young infants, when before surgery the skin was fragile or infected or when there was a seat of infection outside the CNS; the rate of infection was higher when the revised end of the shunt was ventricular or when the operation was a re-insertion after infection. The quality of the post-operative wound was also of great importance. The knowledge of these factors and their elimination whenever possible allowed a new reduction of our infection rate to 4.6%.

Since it had been demonstrated in orthopaedic surgery[5] that prophylactic antibiotic therapy was efficient (allowing a reduction of the infection rate from 3.3% down to 0.9% in 2137 hip arthroplasties) we have used a combination of lincomycin and gentamycin, given at the time of surgery. This antibioitic therapy did not change the rate of infection. We now give cefotaxime and fosfomycine. The rate of infection is lowered to 2.6%, but the number of operations is too small so that this reduction is not at the moment statistically significant. It is not unlikely that the efficiency of prophylactic antibiotic therapy depends on which antibiotics are given. Another possibility might be that prophylatic antibiotic therapy is useless when surgery is performed in a hypersterile environment, as is the case with a surgical isolator.

B) As already stated shunt failures are, over the years, frequent. These failures are the result of mechanical complications such as occlusion, shunt fracture, migration; they can also be the consequence of an improper placement. Finally they are in some cases the consequence of overdrainage: slit ventricles, post-shunt pericerebral collections, craniostenoses. Reviewing the series of "Les Enfants Malades" and the series of "The Sick Children hospital" in Toronto[33] we have been able to specify the characteristics of a shunt which would minimize the rate of these failures: a one piece system, non-flanged ventricular catheter, proximal non-slit valve, open ended distal tubing.

C) However overdrainage is one of the most important problems to solve. Differential pressure valves overdrain always in the upright position. Since

$$F = \frac{DP}{R}$$

(F = flow through the shunt – R = shunt resistance – DP = pressure difference between inlet and outlet) in the prone position, since H (H = height between inlet and outlet of the shunt) should be added to DP in the upright position, it results that, in this position, F increases; F exceeds the CSF secretion rate. This rate being constant, fluid is removed from the venticular reserve in the begining; when this is no longer possible, F decrease while intra-cranial pressure (which is the inlet pressure) becomes negative, favouring pericerebral collections[11] or craniostenoses. When the parenchymal compliance is high, overdrainage results in slit ventricles. Slit ventricles may sometimes be responsible for the "slit ventricle syndrome"; more often slit ventricles favour ventricular catheter obstruction[33]. Since DP varies with the postition of the patient, it was necessary, in order to avoid overdrainage, to design a shunt with a variable resistance (Fig. 5). If DP and R vary simultaneously in the same direction, their ratio, i.e. the flow through the shunt, remains constant. The differential pressure shunt becomes a flow regulator. We have preferred this solution[32] to the antisiphon device designed by Portnoy[27] which is efficient but carries in our opinion a risk of underdrainage in the upright position[20, 32]. The Orbis Sigma Valve acts as a differential pressure valve when the flow is smaller than the CSF secretion. When the flow reaches the value of the CSF secretion, i.e. about 20 to 30 ml per hour, the resistance of the valve increases abruptly so that F

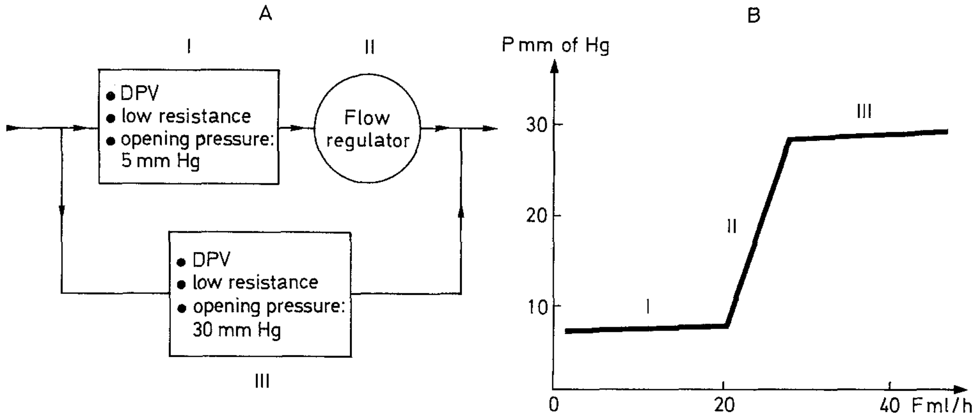

Fig. 5. Pressure-flow curve of the orbis sigma valve. When the flow reaches the normal value of the CSF secretion, the resistance (*i.e.* the slope of the curve) increases so that the differential pressure valve becomes a flow regulator

remains constant even if DP increases. However when DP is above 300 mm H20, the shunt becomes again a differential pressure system; a rapid CSF flow rate is induced, allowing a stabilization of the intracranial pressure. This valve is thus a tree-stage valve system. The first stage allows an appropriate CSF drainage in the prone position. The second stage avoids or at least minimizes overdrainage in the upright position; the third stage is a kind of safety valve.

The comparison of the actuarial risks of shunt malfunction observed before and after the introduction of the Sigma Valve shows that this risk has been reduced two fold (actuarial risk three years after surgery with DP shunts in the Toronto-Paris series 40%, with the Orbis Sigma Valve in 219 shunts inserted in "Les Enfants Malades": 16%). Thus these different attempts have certainly reduced the rate of complications after shunt insertion; they may have also slightly improved the outcome. However the outcome is the clinical expression of the cerebral lesions. These lesions are due to the dilatation of the ventricles before surgery and to the aetiology of the disease. Therefore, in the future, surgery, when indicated, should be performed as early as possible, probably earlier than nowadays; knowledge and prevention of the causes of hydrocephalus should be improved.

References

1. Amacher AL, Wellington J (1984) Infantile hydrocephalus: Long-term results of surgical therapy. Childs Brain 11: 217–229
2. Dandy WE, Blackfan KD (1914) Internal hydrocephalus: An experimental, clinical and pathological study. Am J Dis Child 8: 406–482
3. Dandy WE (1918) Extirpation of the choroid plexus of the lateral ventricle in communicating hydrocephalus. Am J Surg 68: 569
4. Davidoff L (1948) Hydrocephalus and hydrocephalus with meningocele: their treatment by choroid plexectomy. Surg Clin North Am 28: 416
5. Evrard J, Mazas F, Flamant R, Acar J et les members du GETPIA (1981) L'antibiothérapie préventive en chirugie orthopédique. SO.F.C.O.T., Réunion Annuelle, Novembre 1980. Rev Chir Orthop [Suppl. II] 67: 46–59
6. Guiot G, Rougerie J, Fourestier M, Fournier A, Cornay A, Vulmiere J, Graux R (1963) Explorations endoscopiques intracrâniennes. Presse Med 71: 1224–1228
7. Guiot G, Derome P, Hertzog E, Bamberger C, Akerman M (1968) Ventriculo-cisternostomie sous contrôle radioscopique pour hydrocéphalie obstructive. Presse Med 40: 1923–1926
8. Hagberg B (1962) The sequelae of spontaneously arrested infantile hydrocephalus. Develop Med Child Neurol 4: 583–587
9. Hirsch JF, Rénier D, Pierre-Kahn A (1978) Influence of the use of a surgical isolator on the rate of infection in the treatment of hydrocephalus. Childs Brain 4: 137–150
10. Hirsch JF, Hirsch E, Sainte-Rose C, Rénier D, Pierre-Kahn A (1986) Stenosis of the aqueduct of Sylvius. Etiology and treatment. J Neurol Sci 13: 29–39
11. Hoppe-Hirsch E, Sainte-Rose C, Rénier D (1987) Pericerebral collections after shunting. Childs Nerv Syst 3: 97–102
12. Jaksche H, Loew F (1986) Burr hole third ventriculostomy. An unpopular but effective procedure for treatment of certain forms of occlusive hydrocephalus. Acta Neurochir (Wien) 79: 48–51
13. Jansen J (1985) A retrospective analysis 21 to 35 years after birth of hydrocephalic patients born from 1946 to 1955. An overall description of the material and of the criteria used. Acta Neurol Scand 71: 436–447
14. Jansen J, Jorgensen M (1986) Prognostic significance of signs and symptoms in hydrocephalus. Analysis of survival. Acta Neurol Scand 73: 55–65
15. Keucher TR, Mealey J (1979) Long-term results after ventriculoatrial and ventriculoperitoneal shunting for infantile hydrocephalus. J Neurosurg 50: 179–186
16. Lapras C, Poirier N, Deruty R, Bret P, Loyeux O (1975) Le cathetérisme de l'aqueduc de Sylvius. Sa place actuelle dans le traitement chirurgical des sténoses de l'aqueduc de Sylvius, des tumeurs de la fosse cérébrale postérieure et de la syringomyélie. Neurochirurgie 21: 101–109
17. Laurence KH, Coates S (1962) The natural history of hydrocephalus. Detailed analysis of 182 unoperated cases. Arch Dis Child 37: 345–362
18. Laurence KH, Coates S (1967) Spontaneously arrested hydrocephalus results of the re-examinations of 82 survivors from a

series of 182 unoperated cases. Dev Med Child Neurol Suppl 13: 4–13

19. Lorber J, Zachary RB (1968) Primary Congenital Hydrocephalus. Long-term results of controlled therapeutic trial. Arch Dis Child 43: 516–527
20. McCullough DC, Complications with antisiphon devices in hydrocephalics with ventriculo peritoneal shunts in Concepts in Paediatric Neurosurgery II. Karger, Basel, pp 63–74
21. McNickle MF (1947) The surgical treatment of hydrocephalus. A simple method of performing third ventriculostomy. Br J Surg 34: 302–307
22. Matson DD (1949) A new operation for the treatment of communicating hydrocephalus. Report of a case secondary to generalized meningitis. J Neurosurg 6: 238
23. Mazza C, Pasqualin A, Da Pian R (1980) Results of treatment with ventriculo-atrial and ventriculo-peritoneal shunts in infantile nontumoral hydrocephalus. Childs Brain 7: 1–14
24. Mixter WJ (1923) Ventriculostomy and puncture of the floor of the IIIrd ventricle. Preliminary report of a case. Boston Med Surg J 188: 277–278
25. Nulsen FE, Spitz EB (1952) Treatment of hydrocephalus by direct shunt from ventricle to jugular vein. Surg Forum 2: 399
26. Pierre-Kahn A, Rénier D, Bombois B, Askienasy S, Moreau R, Hirsch JF (1975) Place de la ventriculo-cisternostomie dans le traitement des hydrocéphalies non-communicantes. Neurochirurgie 21: 557–559
27. Portnoy HD, Schulte RR, Fox JL (1973) Anti-siphon and reversible occlusion valves for shunting in hydrocephalus and preventing post-shunt subdural haematomas. J Neurosurg 38: 729–738
28. Pudenz RM, Russel FE, Hurd AM, Sheldon CM (1957) Ventriculo-auriculostomy. A technique for shunting cerebrospinal fluid into the right auricle. Preliminary report. J Neurosurg 14: 171–179
29. Putman TJ (1934) Treatment of hydrocephalus by endoscopic coagulation of choroid plexuses: Description of a new instrument. New Engl J Med 210: 1373
30. Rénier D, Lacombe J, Pierre-Kahn A, Sainte-Rose C, Hirsch (1984) The factors of acute shunt infection: Computer analysis of 1174 operations. J Neurosurg 61: 1072–1078
31. Sachs E (1942) Hydrocephalus: Analysis of ninety-eight cases. J Mt Sinai Hosp 9: 767
32. Sainte-Rose C, Hooven M, Hirsch JF (1987) A new approach in the treatment of hydrocephalus. J Neurosurg 66: 213–226
33. Sainte Rose C, Hoffman HJ, Hirsch JF (1989) Shunt failure. In: Marlin AE (ed) Concepts in Pediatric Neurosurgery, Vol 9. Basel, Karger, pp 7–20
34. Scarff JE (1936) Endoscopic treatment of hydrocephalus: Description of a ventriculoscope and preliminary report of cases. Arch Neurol Psychiatry 38: 853–886
35. Scarff JE (1942) Nonobstructive hydrocephalus. Treatment by endoscopic cauterization of choroid plexus. Am J Dis Child 63: 287
36. Scarff JE (1959) Treatment of nonobstructive (communicating) hydrocephalus by cauterization of choroid plexuses: long-term follow-up study. Acta Psychiatr Scand 34: 354
37. Torkildsen A (1939) A new palliative operation in cases of inoperable occlusion of the Sylvian aqueduct. Acta Chir Scand 82: 117–124
38. Yashon D (1963) Prognosis in infantile hydrocephalus: past and present. J Neurosurg 20: 105–111

Correspondence and Reprints: Prof. Jean-Francois Hirsch, Service de Neurochirurgie Pédiatrique, Hôpital Necker Enfants Malades, 149, rue de Sèvres, 75730 Paris, Codex 15, France.