Acta Paediatrica Academiae Scientiarum Hungaricae, Vol. 15 (3—4), pp. 205—209 (1974)

Study of Operated Hydrocephalus Patients Three Years after Surgery

By

E. Paraicz, L. Vékássy and Susan Kovács

Institute of Neurosurgery, Budapest

(Received February 10, 1974)

The development of hydrocephalic children who had been subjected to atrial shunt operation in infancy, has been studied at the age of 3 years. Psychological investigation was carried out in 24 of the 27 surviving cases. Mental development was normal in 14 children, while 10 displayed mental retardation, of whom 8 are still suited for schooling. The grave aetiologic factors are considered primarily responsible for retardation. The functional development achievable by successful surgical treatment is discussed.

Treatment of the hypertensive progressive form of infantile hydrocephalus by ventriculo-atrial shunt has ensured favourable results as to survival. This has made possible to follow-up the patients' somatic and psychic development [8]. A study has therefore been devoted to this question and to the problem whether this kind of shunt meant some advantage over the former ones and as compared to a group of stationary hydrocephalics. While survival after atrial shunting is estimated at 70% [7, 10], data concerning the rate of spontaneous compensation are divergent and those concerning psychic development of operated and non-operated groups are difficult to compare. The rate of survival of non-operated patients amounts to 22% [1], according to other authors, to 33% [3]. Laurence [4] reported on spontaneous compensation in 46% and emphasized that death of the rest had been caused by progression of hydrocephalic hypertension. Of 79 newborns with congenital hydrocephalus observed by Mealey et al. [6] 27 cases have not been operated upon; of these only two survived. On the other hand 34 patients in whom atrial shunt had been performed were alive 7 years later; of these 5 were of normal intelligence and 5 others could also be schooled. In the patients displaying spontaneous compensation, the IQ amounted to 1—0.85 in 41% [4] and in another material in 50% of the cases [2]. In a hypertensive, progressive group of patients subjected to operation, Hemmer and Dill [3] 5 to 10 years later found an IQ of 90—110 in 42% and one of 70—90 in 35% and, in a similar material, Unger et al. [9] found a normal IQ in 60%. Lorber [5] distinguishes a separate group of hydrocephalics with spina bifida; here

spontaneous compensation is more frequent and psychic retardation is less frequent than in hydrocephalics with myelocele who had been operated upon. On the other hand, from among 28 infants with hydrocephalus without spina bifida, whose pallium was less than 10 mm, 22 recovered, and at the age of 7 to 11 years, the IQ of 3 patients amounted to 120—129, it was near the average level in 11, subnormal in 4, and only four were retarded.

Comparison of psychic capacities in the different groups is difficult and serves no purpose since it is obvious that the material falls into two different groups. To the number of cases with spontaneous compensation, i.e. the infants with mild hydrocephalus developing satisfactorily, we must add those who could be kept alive by surgery and who subsequently displayed a satisfactory development of their capacities.

Material

During the 6-year period 1967 to 1973, 173 patients were treated with atrial shunt. Of these, 43 cases have been analyzed in detail. All these patients were about 3 years of age. This seemed important as the aim was to form an opinion of their psychic development in the first place, and 3 years seemed the earliest age suitable.

Mortality and Survival

The most frequent pathogen was *Staphylococcus aureus*. The infection ran an acute course in 6 cases, lasted 6 to 12 weeks in 3 patients, and was chronic in one. Although meningitis prior to surgery might have played a role in the development of hydrocephalus, several such cases have been operated upon without subsequent complication. Necropsy revealed in 5 cases beside inflammatory phenomena an agenesis of the corpus callosum, synventriculia in several cases of ventricular dilatation associated in one infant with microgyria, stenosis of the aqueduct and syringomyelia, and in another one with agenesis of 3 lobes and porencephalia. Thus, in part of the dead such changes would probably have limited the prognosis had they survived.

Number of patients operated upon	43
Alive	27
Dead	16
Cause of death	
Septic complication, septicaemia	10
Cerebrospinal haemorrhage	2
Intercurrent disease	1
Local complication, fistula	1
Unknown	2

Psychological Investigations

Of the 27 survivors, 24 patients were investigated psychologically. Their mean age was 39.2 months. The results were as follows.

(i) *Development*

Sitting: the babies sat up alone at 9.5 months. Standing: standing without support occurred at 12 months. Motor: walking independently at 24 months. Speech: prattling at 7 months,

use of monosyllabic words at 11 months, put words together at 18 months, creation of contact, communicating thoughts, at 26 months.

Several children displayed articulation defects dyslalic in character.

Training: scattering in this respect was wide. The average was trained by the second half of the third year, but several children failed completely.

Dressing: 11 children could not dress by themselves, 9 could do so with help, and only 3 independently.

Eating: One child failed to learn to eat without assistance, 3 eat independently, 30 children eat solid food and use cutlery.

Lateral dominance: for determination of the dominating side, the child was made to draw, etc., by looking through a hole made in a sheet of paper. On this basis, 14 children were right-handed, 6 left-handed and 3 displayed bilaterality.

Drawing: the children had to draw simple forms after a draft (circle, cube, triangle, line). The result was utilized partly for completing the results of the intelligence-test, partly to determine dominance. No specific drawing-mark characteristic of hydrocephalus has been found.

Motor functions: the children were examined in different positions, and during eating and play, they were made to clasp hands and play ball, to perform complex tasks (opening of door, etc.). Opinion of parents and of kindergarten nurses was also taken into consideration. On this basis, 7 patients were lagging behind the expected level. Besides, a certain slowness of movements and uncertain aiming seemed characteristic of hydrocephalic children.

(ii) *Intelligence-test*

In Brunet—Lezin's test, among 24 children the intelligence of 14 corresponded to their age and production was complying with the requirements. Their conduct of life seemed to be ensured, they may learn successfully and adapt themselves socially. Eight children were mentally retarded; the degree of retardation was ½ year in 1 case, 1 year in 5 cases, and 2 in 2 cases. Thuse, 7 children will have to be educated in auxiliary school. The remaining 2 children were oligophrenic.

(iii) *Environment*

The hydrocephalic children were unstable, oversensitive, became suddenly active and even impulsive, could be calmed down with difficulty. In contrast, some were completely passive. The said features were characteristic of both the mentally intact and the retarded patients.

Aetiological and Clinical Data

Aetiologic factors, i.e. those influencing later functions, have been considered separately in the retarded and non-retarded groups.

The experience that in hydrocephaics with spina bifida, mental development is more often normal than in the case of other forms, agrees with data in the literature. In small pre-

Not retarded: 14		Retarded: 10	
Spina bifida (operated on first day of life)	3	Spina bifida	1
Aqueduct occlusion	1	Aqueduct occlusion	1
Purulent meningitis, occlusion	1	Ventriculitis, occlusion	1
Birth in asphyxia	2	Birth in asphyxia	1
Late surgery	1	Late surgery	3
		Toxaemic pregnancy and neonatal subarachnoideal haemorrhage	1
		Preterm birth (1200 and 1900 g)	2
		Post-term delivery	1

matures with a well-functioning shunt, mental retardation is more frequent. The fact that after late operation more infants will be retarded shows that good mental development is best ensured by early surgery.

Except in the prematures, body weight and height complied with the mean corresponding to age.

Mean head circumference was 51.3 cm, i.e. 1 to 1.5 cm more than the mean for the age.

The gravest clinical symptoms have been observed in the two idiots; besides, both were amaurotic. One of them was born at 7 months with 1700 g. The mother had stayed in infected surroundings in the 4th month of pregnancy. At preoperative transillumination, the infant's whole skull was transparent and the fontanel measured 8 cm in diameter. The other patient was born after a toxaemic pregnancy at 8 months, and was treated for subarachnoideal haemorrhage for 2 weeks; when the fontanel was tense, the newborn vomited and had sensorial disturbances.

Neurological symptoms. Five patients suffered from abducens paresis. Its occurrence was independent of psychic development and also of the earlier presence and duration of raised intracranial pressure. However, when the shunt had occluded and reoperation was carried out late, lasting abducens palsy occurred. One of the myelodysplasic patients had a slight spastic palsy, two were incontinent.

Discussion

A minor part of the patients with hydrocephalus is capable of spontaneous correction. This milder form of the disease, recovering essentially without treatment, occurs especially with myelodysplasic hydrocephalus. In this form of the disease treated without surgery, favourable psychic results are frequent, although at most in half of the cases.

Infants with the more severe hypertensive, progressive form of the disease who are the majority and have died formerly, can now be saved by atrial shunt in about 70%. As to their mental development, the reports are striking in that good mental development in this group of patients sur-

passes 50%, and amounted in our material to 70%.

The question arises how to interpret the better late functional result in the more severe form of the disease? The obvious explanation is that in cases with spontaneous compensation the hypertensive condition had to be endured longer than in the operated ones. This too speaks for an early intervention. The principle cannot, however, be followed in every instance. For example, due to the increased danger of septic complications, surgery has to be postponed after a meningo-encephalitis. On the other hand, if a spina bifida is associated with hypertensive hydrocephalus, we have to ascertain whether the case will not compensate spontaneously when the results are better than in operated patients. Still, surgery carried out at an early date remains the way to achieve the best functional results.

From the point of view of late functions, preoperative events, the aetiologic factors of hydrocephalus in which several grave organic diseases have a role, are essentially of the greatest importance. The problems connected with surgery play a role mainly in survival. Since the early solution of occluded shunts may prevent neurological damage, for instance abducens paresis, it is assumed to have a similar role in mental development.

Dr. E. Paraicz
Amerikai út 57
H—1145 Budapest, Hungary

References

1. Foltz, E. L., Shurtleff, D. B.: Five-year comparative study of hydrocephalus in children with and without operation (113 cases). J. Neurosurg. **20,** 1064 (1963).
2. Hagberg, B., Sjögren, I.: The chronic brain syndrome of infantile hydrocephalus. Amer. J. Dis. Child. **112,** 189 (1966).
3. Hemmer, R., Dill, J.: Das Schicksal Hydrocephalus-operierter Kinder. Katamnesen über die körperliche und geistige Entwicklung 5 bis 10 Jahre nach Anlage einer Seitenventrikel-Herzdrainage. Dtsch. med. Wschr. **1,** 1149 (1971).
4. Laurence, K. M.: The natural history of hydrocephalus. Lancet **1,** 1152 (1958).
5. Lorber, J.: Medical and surgical aspects in the treatment of congenital hydrocephalus. Neuropädiatrie **3,** 239 (1971).
6. Mealey, J. Jr., Gilmore, R. L., Bubb, M. O.: The prognosis of hydrocephalus overt at birth. J. Neurosurg. **39,** 348 (1973).
7. Nulsen, F. E.: Discussion. J. Neurosurg. **20,** 1078 (1963).
8. Paraicz, E., Katona, F., Szénásy, J.: Adatok a csecsemőkori hydrocephalus kezelésének eredményeiről és néhány tényező szerepéről ezek kialakításában. Gyermekgyógyászat **23,** 475 (1972).
9. Unger, R. R., Ollmann, S., Schmitz, F., Görner, W.: Neurologisch-psychiatrische Nachuntersuchungsergebnisse nach Shuntoperationen (Spitz—Holter) wegen frühkindlichen Hydrocephalus. Dtsch. Gesundh. Wes. **24,** 1427 (1969).
10. Yashon, D.: Prognosis in infantile hydrocephalus. J. Neurosurg. **20,** 105 (1963).