

Adult quality of life in congenital hydrocephalus operated cases. A twenty years retrospective study

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Abstract: Congenital hydrocephalus is a health problem in many countries and in Romania the pediatric neurosurgical department of the Emergency Hospital “Bagdasar-Arseni” has a large number of such patients. This is a retrospective study and it includes the patients with congenital hydrocephalus operated between 1992 and 2012 in the pediatric neurosurgical department of the Emergency Hospital “Bagdasar-Arseni”. The functional outcome was assessed using Karnofsky Performance Scale, Hydrocephalus Outcome Questionnaire and Glasgow outcome scale. The total number of the patients was 372, with a predominance of boys (212 boys versus 160 girls) and at the time of our study 168 patients were over 16 years old. Functional outcome of the children over 16 years old assessed using Karnofsky Performance Scale, showed that 73 patients were above 80 and leading independent lives, and 95 were less than 80 points. The results would be better if all these patients would benefit from schooling for children with special needs.

Key words: congenital hydrocephalus, quality of life, retrospective study.

Introduction

Congenital hydrocephalus is a health problem in many countries, with an incidence of 0,2 to 0,8/1000 live births in USA. The techniques of shunting have permanently evolved since the early days of Holter. They became routine in many pediatric neurosurgical departments but the data that describe mortality and morbidity and that certify the improvement in the quality of life of these patients are a few. The old saying: “once a shunt, always a shunt” hasn’t, till now, been completely contradicted. Even further, data regarding the remote prognosis of these children that reach adulthood are even less.

Accordingly, clinical scores have been designed to assess the functional and vital outcome of the children and adults that have been operated in childhood for congenital

hydrocephalus. There is no definite consensus over the best scale to use, making comparison between different series difficult.

Material and methods

For many years the pediatric neurosurgical department of the Emergency Hospital “Bagdasar-Arseni” was the only unit in Romania that could take in charge children with congenital hydrocephalus. Also, the children who were fortunate to become adults were treated in the same hospital. The study that we have designed is a retrospective one. There were selected the patients with congenital hydrocephalus without any other pathology, operated between 1992 and 2012 in the pediatric neurosurgical department of the Emergency Hospital “Bagdasar-Arseni”. The

follow-up of the patients is a life-long one and patients were invited to check-up once a year. The hospital protocol is to revise any shunt that has been ruptured, has become short, or otherwise nonfunctional, provided that the patient is dependent on the shunt. The functional outcome was assessed using Karnofsky Performance Scale, Hydrocephalus Outcome Questionnaire and Glasgow outcome scale. On the Karnofsky scale, 100 is asymptomatic, 80 able to assume normal life, 0= dead. With Glasgow Coma Scale, according to the WFNS, 1 is normal life and 5 is dead. Schooling was appreciated as normal, school with special help or none at all. Social status depended on the employment or non-employment of the patients.

Results

The total number of the patients was 372, with a predominance of boys (212 boys versus 160 girls). The reoperation was due to shunt obstruction and infection. There were 1190 reoperations with an average of 3,19 reoperations for a patient. In 68 patients, reoperation was made for shunt infection with an average of 3 reoperations for a patient. A minimum of 2 operations and a maximum of 23 operations for a patient were performed. Only 10 patients were without sequelae. A large number, 131 patients had motor deficits and even more, 245, had cognitive deficits. Unfortunately, 31 patients died. At the time of our study, 168 patients were over 16 years old. No one of our patients gained independency from their shunts.

As we can see from the graphic, for the children operated when they were under 1 year of age, first reoperation was mainly between 4 and 7 years of age (162 cases).

Functional outcome of the children over 16 years old assessed using Karnofsky Performance Scale, showed that 73 patients were above 80 and leading independent lives, and 95 were less than 80 points.

Concerning schooling, there was a disappointing 27,1% (101) of the children that followed normal school, with 12,9% (48) of the children who could not attend any school. As for the rest, 223 children (59,9%) needed special schooling.

Unemployment in the group of 168 patients that were over 16 years old, was pretty high, of over 50%, with patients that over the years had a job and had lost it, and patients that have never worked.

Discussion

It is well known that shunt malfunction is a permanent risk for the hydrocephalic patients that were treated with shunts. Earlier the shunt, worse the prognosis. As we have seen, the reoperation for different reasons for shunt malfunction, is greater between 4 and 7 years old and then, between 8 and 11 years old, if the children were shunted before age of 1 year old. As for the children who were treated with ETV (5 cases), the number is too small to draw a conclusion, but it seems that an obstruction of the stomy is more dangerous than a merely shunt obstruction, as acute hydrocephalus leads to coma and impending death.

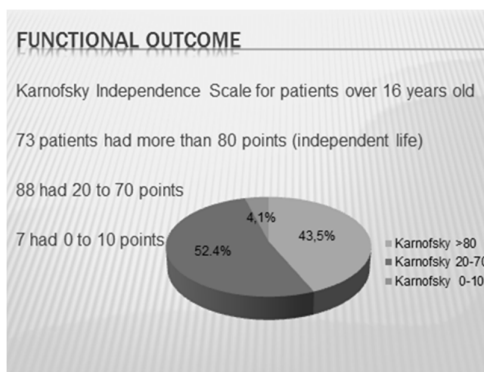
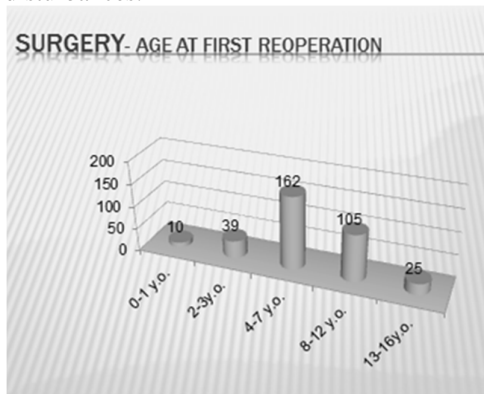
Concerning the shunt infection, the most frequent germ was *Staphylococcus epidermidis*, which means that proper care is mandatory for these patients. Infection of the shunt is usually heralded by the appearance on the abdominal ultrasound of a peritoneal cyst which precludes the normal absorption of the CSF.

A special problem is that of an asymptomatic patient that has been shunted for years and presents with a ruptured shunt. There are 2 ways to deal with. The first is to revise the shunt, and even there is a large gap between the stumps of the shunt, and the surgeon sees only one CSF drop drooling from the proximal stump, than the shunt is to be considered needed and the gap should be repaired. The second is to ligate the shunt at the neck of the patient and see if he would be

asymptomatic. That is a dangerous maneuver, because the shunt might be in fact functional and the patient would be in deep coma in short time. We should stress, as we have already said, that shunt independence is rather exceptional.

Mortality is a fact that cannot be overlooked. 31 of our patients died of shunt malfunction, mostly because of obstruction of the shunt. This is why the close follow-up of the shunts is the only way to prevent such an occurrence.

Speaking about sequelae, the cause of hydrocephalus and frequent shunt malfunctions are the main causes of neurological sequelae and psychological disturbances.



Conclusions

Prolongation of life span, with or without

sequelae, depending on a shunt, is usually all that we could do for these patients. Life-long follow-up is the only way to prevent shunt malfunction and sudden death and this is the work of a dedicated.

The education system should provide a repeated psychological evaluation and a system schooling for children with special needs.

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