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Short communication



# Fatal Shunt Failure 40 Years after Implantation of a Lumbouretral Shunt in a Hydrocephalic Infant

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## Abstract

Long-term results in hydrocephalic children treated with CSF diversion to the ureter have not been reported. We present our experience with seven children shunted with lumbo-uretral shunts during the years 1957-1959. They presented as infants (2-9 months old) with severe head enlargement (46 – 61 cm), all of them with very tense heads and sunset signs. Six of the patients died within two years. The last patient grew up and had been in full-time work for 23 years, when he unexpectedly died from acute shunt failure. He had not been at regular follow-up since his early years and had never been to a CT until his dramatic shunt failure. Although shunt failure may occur after decades with good function of the shunt, the topic is troublesome since very long-time follow-up together with patient awareness may seem appropriate to avoid or reduce the risk of future shunt failure.

**Abbreviations:** CSF: Cerebrospinal Fluid; LU Shunt: Lumbo-Uretral Shunt; IIH: Idiopathic Intracranial Hypertension; VA shunt: Ventriculoatrial Shunt; VCS: Ventriculocisternostomy; VP Shunt: Ventriculoperitoneal Shunt

## Introduction

In 1938 Walter Dandy published a series of three infants treated surgically for severe communicating hydrocephalus [1]. He performed electrocauterization of plexus choroideus in the lateral ventricles as well as in the fourth ventricle. One of the children died within short time, but in the other two, the head circumference stabilized, and they appeared to do quite well during the next year or two. Arne Torkildsen introduced ventriculocisternostomy (VCS) for non-communicating hydrocephalus in 1939 [2,3]. Management of severe hydrocephalus in small children was, however, a great challenge to neurosurgeons during the 1940ies, since VCS was not found applicable in infants and most of them had communicating HC. Donald Matson introduced CSF diversion to the ureter after nephrectomy from the lumbar dural sac in 1949 [4]. In selected cases this procedure was in use for the next decade, until modern VA and VP shunts took over [5-7]. In our department lumbo-uretral shunting including nephrectomy was considered a major procedure in small babies and the method was in use only during the years 1957 to 1959.

## Results

Seven infants aged 2-9 months presented with clinical signs and symptoms of increased ICP (rapidly growing and tense heads, sunset signs).The head circumference varied from 46 to 61 cm and ventriculography demonstrated severely enlarged ventricles. In three of the children the posterior fossa was opened, and in two of these a ventrculocisternostomia (Torkildsen procedure) was performed but was not successful in relieving the high ICP.

All patients underwent a CSF diversion procedure to the ureter after nephrectomy. The proximal catheter was introduced in the lateral ventricle in one patient and in the lumbar dural sac in the other six. In all these infants, the drainage was initially successful (reduced head circumference and tension). All patients were

given 2 g extra salt daily. At 5 weeks follow-up, the clinical effect had disappeared in one patient due to shunt failure that resolved after shunt revision (blockage the proximal catheter, successful after flushing). Six of the patients died within 2 years, one in the early postoperative period, and one 6 months after surgery due to meningitis.

The last infant was a 4-months-old boy who head circumference of 51 cm, who underwent a LU- shunt procedure in May 1957. The shunt appeared to work well shortly after operation, as well as at one- year follow-up. Thereafter, he grew up and finished school without major problems. He was in full-time work from the age of 17 years until his sudden death more than 40 years after shunt implantation.

In January 1998, after being to work as usual 12 hours earlier, he was found semi-comatose, and was brought to the local hospital. The pupils were semi-dilated with only slight reaction to light. He was vomiting and responded with simple words and had bilateral motor responses. The blood pressure was 170/130 mm Hg. The head circumference was 60 cm. A CT demonstrated ventriculomegaly, and he was transferred to the neurosurgical department, hyperventilated and under Mannitol infusion with small reactive pupils. A VP shunt was immediately implanted, and the CSF pressure was found extremely high in spite of hyperventilation. He did not wake-up and demonstrated left-sided adverse motor response, with flexion movements on the right side during the next days. Repeat CT demonstrated infarction of the right hemisphere. He died in the local hospital six days after the onset. We believe that his death was caused by CSF obstruction leading to secondary herniation.

## Discussion

Management of infants with severe and progressive HC was a great challenge to neurosurgeons in the 1950ies. Multiple plexus choroideus resections were dangerous and anterior III-ventriculostomy as well as VCS proved to be unsuccessful in small children. Matson and coworkers obtained satisfactory results with VU-shunting in selected patients in terms of survival and clinical development. Problems with salt depletion were well recognized and reported but long-term follow-up results have not been reported.

In retrospect, it was a surprise that one out of our children grew up and was in full-time work for more than two decades, until an unexpected sudden death. After the first few years with additional salt intake, pleasing development during childhood into adulthood followed, and questions like shunt failure or shunt dependence were not raised. This 40-year-old man had never been to a CT of the head until the acute CSF obstruction unexpectedly appeared.

We must recognize that this 40-year survival (without revisions) of an infant treated with LU-shunt in 1957 was unexpected. It is, however, also our opinion that the complete lack of long-term follow-up of this patient raises fundamental questions. We have found that about half of our children who underwent CSF diversion (VA shunts) in the late 1960ies are still alive, and most of them appear to be shunt dependent [8]. We have also seen late shunt dependency and shunt failure in children shunted for IIH [9]. Extremely late shunt failures after many years of well-functioning treatment do occur and can be life-threatening. In the 1950ies permanent shunt dependency and the need for long-term follow-up of shunted individuals had not been established. When these questions have not been addressed for several decades, clinical signs and symptoms of acute shunt failure may come unexpected and be easily misunderstood. Life-long follow-up and/or implanting a patient awareness program may therefor seem appropriate.

## Acknowledgement

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## Conflict of Interest

No conflict of interest

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