Dandy-Walker malformation: Surgical treatment of 17 cases

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Abstract: Seventeen patients with Dandy-Walker malformation were treated between 1985 and 2006. Hydrocephalus was present in 88% of patients at time of diagnosis.

Cystoperitoneal and ventriculoperitoneal shunting were performed as primary modality of treatment in 2 and 13 cases respectively. Nine of them later required an additional shunting through a Y montage. Combined ventriculocystoperitoneal shunting was performed as primary modality in 2 patients.

Following operation, two thirds of the patients had an IQ of 80 or more on long term evaluation and the outcome is even more favourable for children above 3 years. Early and adequate treatment of hydrocephalus is the single most important factor in providing the potential for normal intellectual development

At the present time Dandy-Walker malformation is undoubtfully best managed by combined shunting of the cysts and lateral ventricles as initial procedure. *(p25-30)*

Key words: *Dandy-Walker malformation, hydrocephalus, cerebrospinal fluid shunt and posterior fossa cyst*

Introduction

Dandy-Walker malformation (DWM) is classically described by the neuropathological triad of cystic dilatation of the 4th ventricle, hypoplasia of the cerebellar vermis, and hydrocephalus.[11] Although hydrocephalus occurs in the majority of cases, it is not an aid to the diagnosis of DWM.[3,9,11,13] Like most congenital central nervous system (CNS) anomalies, clinical expression of DWM usually appears early in life, frequently below one year of age.[20,22] The majority of children present with symptoms and signs of hydrocephalus and increased intracranial pressure.[13]

Material and methods

From 1985 to 2006, 38 cases of patients with diagnosed Dandy-Walker malformations were reviewed retrospectively. Seventeen cases were asymptomatic neurologically and therefore treated conservatively. Twenty-one cases were neurologically symptomatic. Four of them were associated with severe systemic anomalies and the parents refused the surgical treatment as the prognosis was terminal. The remaining 17 patients were treated surgically.

Age of patient at the time of diagnosis ranged from 15 days to 4 years (84% of them diagnosed at or before age of one year). There were 7 boys and 10 girls. The common presenting symptoms and signs in these patients were macrocephaly in all patients and gait disturbance in 4 patients. There was no oculomotor disturbances nor cerebellar or brain stem dysfunctions. Hydrocephalus was present in 15 cases (88%). Associated congenital systemic anomalies were present in 7 patients (42%).

The major diagnostic procedure was computed tomography (CT) in 10 cases and magnetic resonance imaging (MRI) in 7 patients. Large posterior fossa, dysgenesis of cerebellar vermis and cystic enlargement of the 4th ventricle were the positive findings recorded in all patients. All our patients had hydrocephalus; mild, in 7 cases and severe in 8 cases. No associated hemispheric or skull anomalies were detected.

Ventriculoperitoneal (VP) shunting was performed as primary modality of treatment in 13 cases, where the posterior fossa cyst was moderate in size and asymptomatic, and the hydrocephalus was predominant clinically and radiologically.

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Seven of them required, after a period of one month, an additional cystoperitoneal shunt through a Y montage as these cysts were large and still compressed elements of the posterior fossa.

***Case 1*:** Dandy-Walker malformation with hydrocephalus; the primary treatment was a ventriculocystoperitoneal shunting through Y connection (Fig. 1).

Cystoperitoneal shunting was performed in 2 patients as primary modality of treatment, as these cysts were large but with mild hydrocephalus. Later, it was changed to ventriculocystoperitoneal shunting through a Y montage as the hydrocephalus, which had diminished initially, reappeared with subsequent neurological deterioration.

***Case 2*:** Dandy-Walker malformation without hydrocephalus; the primary treatment was cystoperitneal shunting. Secondary, converted into ventriculocystoperitoneal shunting through Y connection after complication of active hydrocephalus (Fig. 2).

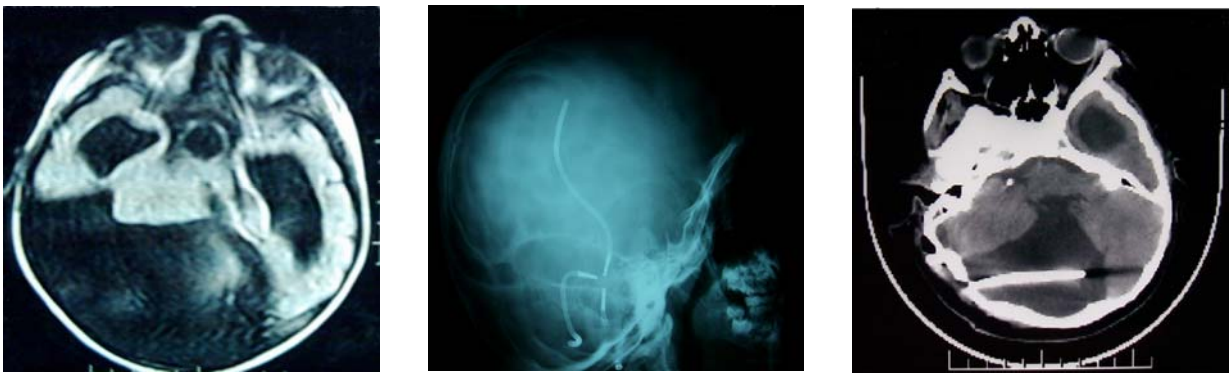


Figure 1 - (**a**) ↑← Preoperative MRI T1 axial view (**b**) ↑ Y connection (**c**) ↑→ Placement of the tube in the posterior fossa cyst

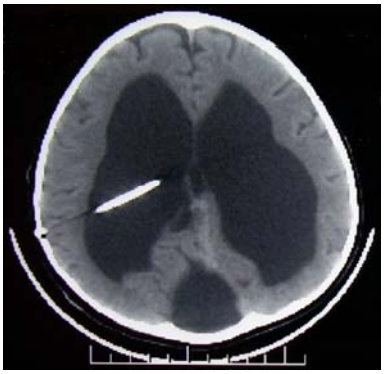
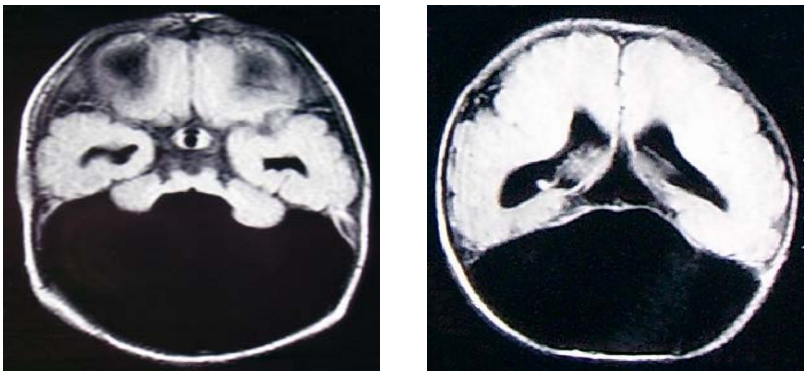


Figure 1d - Placement of the tube in the lateral ventricle



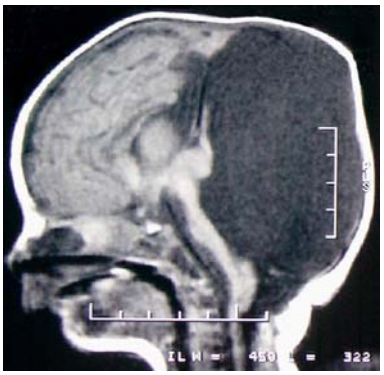
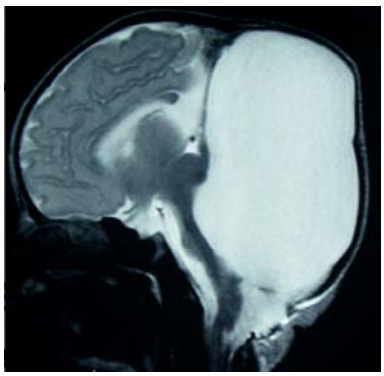


Figure 2 - (**a**) ↑← Preoperative MRI T1 axial view posterior fossa cyst. (**b**) ↑→ Preoperative MRI T1 axial view lateral ventricles. (**c**) ← Preoperative MRI T1 sagittal view. (**d**) → Preoperative MRI T2 sagittal view



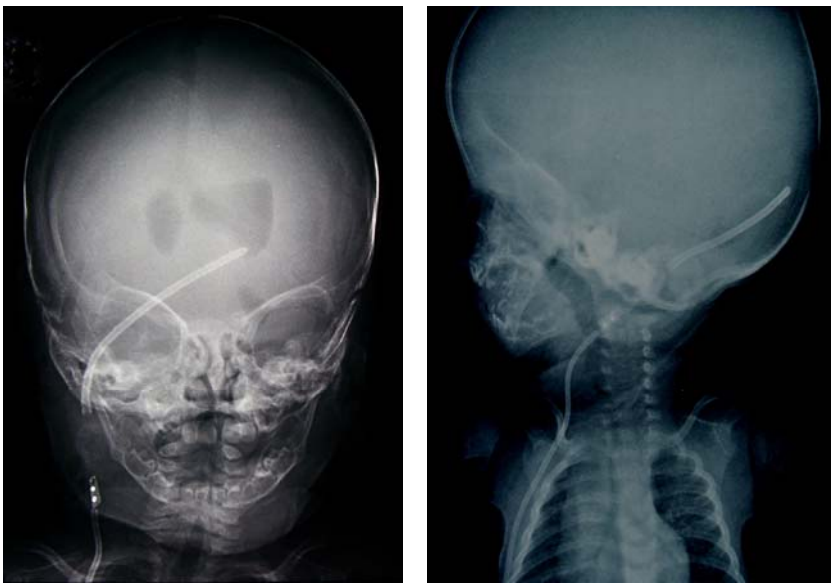


Figure 2 - (e) Cystoperitoneal shunting frontal view. **(f)** Cystoperitoneal shunting lateral view

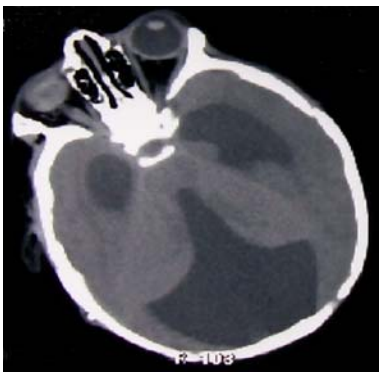


Figure 2g - Preoperative MRI T1 of the posterior fossa

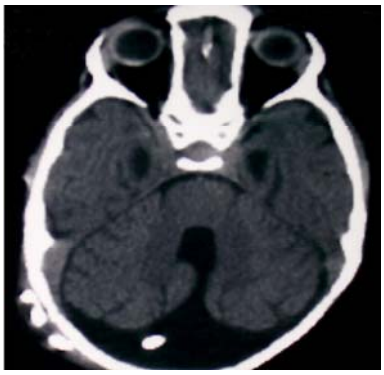


Figure 2h - Postoperative MRI T1 of the posterior fossa

Combined ventriculocystoperitoneal shunting was used as primary modality in 2 patients where hydrocephalus was present and the posterior fossa cyst was large.

No craniotomy was performed with fenestration / excision of the membrane of the posterior fossa cyst.

Low pressure valve was placed in 15 cases and a smart valve was placed in 2 cases.

Revision had to be carried out in one case of cystoperitoneal shunting due to malfunction and 5 for lengthening of the shunt system.

Four cases presented with infection. One of them was lethal due to severe intracranial hypertension and late transfer to the hospital. The others were treated for their infection and successfully surgically managed. There was no subdural collection. One patient died from a severe cardiac deficiency.

Discussion

The origin of DWM is still unclear and remains a source of controversy. It is generally thought that DWM is caused by an atresia of Magendie and Lushcka foramens, as reported by Dandy and Blackfan, and Taggart and Walker.[4,24]

According to Hirsh this theory is untenable.[13] This atresia of the foramina of the 4th ventricle would cause a hydrocephalus that would immediately be obvious at the time of birth. In his series, 80% of the infants are not hydrocephalic at the time of birth.[13] He found an imperforation of Magendie in all 9 cases, approached surgically, and there was communication between the 4th ventricle and the subarachnoid space in 2 of the 9 cases. Of 22 patients studied, 18 (82%) showed a communication between the 4th ventricle and the subarachnoid space (probably through the foramen of Lushka) using air or metrizamide or isotope studies, which proved for us insufficient for normal circulation of cerebrospinal fluid (CSF).

Mechanism of associated hydrocephalus that could be moderate or severe at time of diagnosis is sill unclear. Hirsh reported several cases where DWM was diagnosed as macrocrania due to enlarged posterior fossa. Later these patients developed secondary hydrocephalus.[13] The secondary opening of the foramen of Lusaka, which is supposed to take place at some stage following birth would explain the absence of hydrocephalus at birth. Hirsh explains the delayed indication of hydrocephalus by a possible bleeding that occurs at delivery and ensuing secondary obstruction of the communication between the 4th ventricle and the subarachnoid space.[8] He concludes, therefore, that CSF circulation is not completely blocked and the lack of

balance between CSF secretion and CSF resorption at a normal intracranial pressure develops after birth.[8] The management of DWM has been heavily debated and treatment options range from craniotomy with cyst fenestration or excision to CSF shunting procedures.[8,13]

Lateral ventricle shunting is the first temptation in a hydrocephalic child or new born with DWM, which is an easy procedure and effective in treating hydrocephalus, but shunting of lateral ventricles alone may still fail despite radiographic demonstration of anatomical patency of the aqueduct.[18] In the series where the first modality of treatment was VP shunting an additional derivation of the posterior fossa cyst is unmanageable. Osenbach placed 12 shunting combined derivation systems in 13 VP and 4 cystoperitoneal shunt procedures.[18] Three cases of 10 VP shunts performed by Sawaya shunts were converted to combined ventriculocystoperitoneal shunts.[23]

Kumar, et al in his series converted a total of 8 cases into a combined shunt after 28 simple shunts (VP).[15] Asai, et al encountered similar problems in their series of patients.[1] Nine of 21 patients (43%) initially managed with a lateral ventricular shunt required additional cystoperitoneal shunting. Hirsh performed a single shunting procedure in all cases in his series.[13] None of them were converted to a double shunting. This necessity of combining cystoperitoneal shunt to the VP shunt is due to persistent posterior fossa cyst due to acqueducal stenosis or entrappement of the 4th ventricles.[7,12]

Pillay, et al reported upward transtentorial herniation following a VP shunt in cases of Dandy-Walker cysts.[19] A keyhole configuration of the 4th ventricle seen on axial CT images of patients with Dandy-Walker cysts has been reported to be an indication of upward transtentorial herniation, an entity distinct from entrapped 4th ventricles of other causes, and other posterior fossa cysts which may also develop into upward transtentorial herniation. We did not observe such a complication in our series.

Most authors would currently agree that shunting of the cyst should be performed in almost all cases as this gives direct decompression of the posterior fossa structures thus relieving brainstem and cerebellum in patients symptoms.[19]

There continues to be debate over whether simultaneous shunting of the lateral ventricle should also be carried out. Based on a lack of evidence of stenosis between the supratentorial compartment and the posterior fossa cyst, Sawaya and McLaurin advocated a policy of shunting the cyst alone.[23] These authors reported satisfactory control of hydrocephalus and intracranial hypertension in all patients treated in such a fashion.[22] They felt that the lack of collapse of the cyst around the shunt tubing minimized the risk of malfunction.[22] Hirsch, et al successfully treated 13 children with cystoperitoneal shunts with no apparent complications.[13] The authors felt that equivalent results could be obtained with a simple cystoperitoneal shunt compared to a double shunt system. Asai, et al reviewed 35 children with DWM at the Toronto Hospital for Sick Children of whom 10 were successfully managed with a simple cystoperitoneal shunt.[1] The authors noted improvement in hydrocephalus and a reduction in the posterior fossa cyst in all 10 children and concluded that the ideal treatment for DWM is shunting of the 4th ventricular cyst.

In contrast to upward 4th transtentorial herniation with a VP shunt, Naidich, et al reported that a cystoperitoneal shunt alone leads to chronic cerebral herniation in shunted Dandy-Walker malformations.[17] They found that in patients with Dandy-Walker malformation, the high position of the tentorium, the wide incisura, the hypoplastic cerebellum and the large posterior fossa cyst predispose to chronic downward transincisural herniation of the cerebral hemisphere when the cystoperitoneal shunt reduces infratentorial volume and pressure. Once established, the herniation may persist despite multiple shunt revisions, even after the restoration of normal pressure. We did not observe such a complication in our series.

Maria, et al suggest the utilization of cystoperitoneal shunts when patency of aqueduct can be confirmed.[16] Nowadays, this could be suspected on standard MRI and confirmed by a CSF flow dynamic study to confirm the CSF flow velocity and its direction of flow at aqueductal level which could help in decision making.

Combined shunting of the lateral ventricles and posterior fossa cyst has been advocated by several authors.[10,14,25,26] Udvarhelyi and Epstein as well as Tal, et al recommended a double shunt in all cases in which complete aqueductal obstruction is demonstrated.[25,26] James, et al found that the most successful method of treatment was a combined shunt.[14] All 6 children treated in this fashion showed prompt resolution of their symptoms. Osenbach also favoured a combined ventriculocystoperitoneal shunt as the initial procedure in children with DWM.[18] Combined shunting in his series was judged successful in 11 of 12 (92%) children, initially treated in this fashion based on control of hydrocephalus, reduction of intracranial pressure and reduction in the size of the posterior fossa cyst. The incidence of shunt-related complications was no greater in this group than in those patients who received a single shunt. Some authors argue against using a double shunt citing the potential danger of developing a transtentorial pressure gradient should one shunt fail. However, we have not experienced this problem in our patients.

Membrane excision was initially proposed by Dandy for the treatment of Dandy-Walker syndrome. The results using this form of therapy were poor, with a high failure rate. This surgical approach was based on an assumption that the blocked outlets of the 4th ventricle were responsible for expansion of the 4th ventricle and supratentorial hydrocephalus.[6] Dandy and Blackfan proposed a membrane excision, that fell into disrepute due to a high mortality rate of 10% and a failure rate of 70%.[4]

However, Udvarhelyi and Epstein still advocate this treatment by way of cyst wall excision in children aged over 3 years.[26] Kumar, et al reported no complications following surgery after wide membrane fenestration, proposed as their primary modality in 2 patients over 3 years of age.[15]

Since recent studies with a large number of patients treated surgically are not available it is difficult to comment on the complications and outcome of the above mentioned series results. Comparative randomized large studies are still needed to compare the membrane fenestration results to those of CSF shunting.

In our series we did not perform direct surgical approach of the posterior fossa. We believe that it is a potential source of high risk complications and we preserve it for use in DWM cases uncontrolled by CSF derivation.

The high mortality rate of our series had been greatly reduced due to improvements in paediatric anaesthesia and intensive care, shunting devices, and the elimination of posterior fossa exploration from the surgical armamentarium.

As has been the case with hydrocephalic children in general, improved survival has increased concern regarding intellectual development in these children. The prognosis regarding intellectual development in patients with DWM is variable. Several series have reported subnormal intelligence in up to 50% of children with DWM possibly related to the presence of associated CNS anomalies such as agenesis of the corpus callosum or cortical dysgenesis despite good control of hydrocephalus.[1,6,13,23] Sawaya and McLaurin reported that 71% of children in their series had an IQ < 83; a disappointing figure despite what they felt to be adequate early treatment.[24] They noted no correlation between intellect and other CNS anomalies. Raimondi and Soare noticed that children with DWM had markedly lower mean IQ scores than children with hydrocephalus from other causes.[22] Raimondi, et al in a subsequent series of children with DWM, reported that less than 10% of children tested had a normal IQ.[22] They concluded that the unfavourable outcome in their patients was not indicative of unsuccessful surgical management but was likely related to associated cerebral anomalies. In contrast, Carmel, et al reported more encouraging results with 50% of children having normal intellect at long-term follow-up.[2] They suggested the most critical factor in providing the potential for optimal intellectual development is adequate control of hydrocephalus. In Osenbach series, 35% of the surviving patients were judged to have subnormal intelligence by formal testing.[18] He noted no correlation between intellectual development and CNS anomalies. In our series, two thirds of the patients had an IQ of 80 or more on long term review and the outcome is more favourable for children above 3 years. Obviously, the common agreement that in the absence of major CNS malformations, early and adequate treatment of hydrocephalus is the single most important factor in providing the potential for normal intellectual development.

Conclusion

In our experience it is important for DWM to be understood hydrodynamically and therefore managed skillfully. The CSF flow study on MRI is now of paramount importance to analyze the CSF flow direction in order to better understand and then propose an appropriate treatment. At the present time this entity is undoubtfully best managed by cysto-peritoneal shunting alone, if the hydrocephalus is moderate or combined to a VP shunting through a Y connection or if hydrocephalus is active. These procedures should performed by a skilled and senior neurosurgeon to reduce the possible complications of shunting procedure.

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GENTLE REMINDER

Associated anomalies of the central nervous system in Dandy-Walker syndrome

- Absence of medullary pyramid
- Agyria, aqueductal stenosis
- Cavum septi pellucidi
- Cerebellar folial anomalies
- Chiari malformation Type III, cyst of choroid plexus
- Diverticular cyst of third ventricle
- Dysgenesis of corpus callosum
- Encephalocele, fused hypothalami
- Holoprosencephaly, infundibular hamartoma
- Malformation of inferior olivary nuclei
- Meningocele, microcephaly
- Neurocutaneous melanosis
- Non-specific cerebral gyral anomalies
- Polymicrogyria, specific cerebral gyral anomalies
- Syringomyelia