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The Prognosis of Children with Hydrocephalus and Congenital Heart Disease

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Key Words
Children  •  Hydrocephalus  •  Heart disease

Abstract
The association of congenital hydrocephalus and heart disease in children is infrequent, but may present considerable dilemmas in management. This report describes the treatment and prognosis of 11 children with both clinical problems. There were 5 males and 6 females. Hydrocephalus occurred following aqueductal stenosis in 5 children and the Dandy-Walker malformation in 3. Three children were diagnosed with idiopathic hydrocephalus. Ten children underwent cerebrospinal fluid diversion procedures for control of hydrocephalus. Five children received pharmacological therapy for cardiac disease; 4 children required surgical correction. Two children died from medical conditions; 2 families declined treatment. Follow-up from 2 to 7 years in the remaining 7 children demonstrated moderate or severe neurodevelopmental disability in 5. One child at 2 years of age showed borderline developmental disability while 1 child is developing normally at 10 years of age. Overall the occurrence of symptomatic hydrocephalus and heart disease in the perinatal period resulted in mortality or neurodevelopmental disability in 9/11 children.

Introduction

The prognosis for children with hydrocephalus or congenital heart disease has been reviewed in many articles and texts. For example, infants diagnosed with congenital hydrocephalus have demonstrated variable outcomes, but as high as 60% may be normal on follow-up [1–6]. Risk factors for mortality and poor outcome in these patients included associated central nervous system anomalies and the thickness of the cortical mantle prior to and after treatment. Similar to hydrocephalus, the prognosis in congenital heart disease depends on etiology [7]. Improved diagnostic techniques and medical and surgical therapies have reduced mortality so that contemporary statistics indicate that 85% of these children will survive into adulthood. Improvements in neurological outcomes have been shown, although there are several well-described neurological complications associated with congenital heart disease or its therapy [8–12].

While these reviews addressed outcomes in children with hydrocephalus or congenital heart disease, the uncommon occurrence of both of these potentially treatable conditions in young patients has not been examined in detail. This report will describe the clinical course and neurodevelopmental outcome in 11 children with the association of hydrocephalus and congenital heart disease.
Children with Hydrocephalus and Congenital Heart Disease

<table>
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<tr>
<th>Patient</th>
<th>Age at diagnosis of hydrocephalus</th>
<th>Etiology of hydrocephalus</th>
<th>Etiology of CHD</th>
<th>Surgical treatment</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>N.W.</td>
<td>prenatal</td>
<td>aqueductal stenosis</td>
<td>AVC, coarctation, PDA, AOS</td>
<td>VPS</td>
<td>died 9 days of age</td>
</tr>
<tr>
<td>K.S.</td>
<td>birth</td>
<td>DWM</td>
<td>ASD, VSD, cardiomyopathy</td>
<td>none</td>
<td>died 19 days of age</td>
</tr>
<tr>
<td>T.S.</td>
<td>2 months</td>
<td>idiopathic</td>
<td>corrected transposition</td>
<td>VPS</td>
<td>died 3 months secondary to progressive respiratory failure</td>
</tr>
<tr>
<td>B.C.</td>
<td>prenatal</td>
<td>DWM</td>
<td>AVC</td>
<td>CPS, repair AVC</td>
<td>died 8 months following multi-system organ failure</td>
</tr>
<tr>
<td>T.F.</td>
<td>3 months</td>
<td>aqueductal stenosis</td>
<td>PFO</td>
<td>VPS</td>
<td>severe developmental delay at 2 years of age</td>
</tr>
<tr>
<td>C.C.</td>
<td>prenatal</td>
<td>DWM</td>
<td>ASD, PDA</td>
<td>VPS</td>
<td>severe developmental delay at 3 years of age</td>
</tr>
<tr>
<td>J.F.</td>
<td>2 months</td>
<td>aqueductal stenosis</td>
<td>PDA, ASD</td>
<td>VPS, repair of PDA and ASD</td>
<td>severe developmental delay at 2 years of age</td>
</tr>
<tr>
<td>M.C.</td>
<td>prenatal</td>
<td>idiopathic</td>
<td>small VSD</td>
<td>VPS</td>
<td>moderate developmental delay, seizure disorder at 4 years of age</td>
</tr>
<tr>
<td>R.R.</td>
<td>5 months</td>
<td>idiopathic</td>
<td>small VSD</td>
<td>VPS</td>
<td>moderate developmental delay at 7 years of age</td>
</tr>
<tr>
<td>A.P.</td>
<td>2 months</td>
<td>aqueductal stenosis</td>
<td>coarctation</td>
<td>VPS, repair of coarctation</td>
<td>normal at 10 years of age</td>
</tr>
<tr>
<td>A.K.</td>
<td>3 months</td>
<td>aqueductal stenosis</td>
<td>AVC, VSD, MR, TR, pulmonary HTN</td>
<td>VPS, repair of cardiac anomalies</td>
<td>borderline developmental delay, 2 years of age</td>
</tr>
</tbody>
</table>

AOS = Aortic stenosis; ASD = atrial septal defect; AVC = arteriovenous canal; CHD = congenital heart disease; HTN = hypertension; MR = mitral valve regurgitation; PDA = patent ductus arteriosis; PFO = patent foramen ovale; TR = tricuspid valve regurgitation; VSD = ventricular septal defect.

Methods

The records of the Children’s Hospital of Illinois at Saint Francis Medical Center, a tertiary referral center for pediatric neurosurgery and cardiac surgery, were reviewed from 1987 to 1997. Children were excluded if the hydrocephalus followed intracranial hemorrhage or infection. Hydrocephalus was diagnosed by either CT or MRI scan. The structures of the cardiac anomalies were assessed by echocardiogram, angiography or, in 1 case, at autopsy.

Children were followed from time of treatment or diagnosis. Neurodevelopmental outcome was assessed by either a developmental pediatrician or the staff of an early intervention program and were classified as normal, borderline, moderate or severe delay [13].

Results

Two children developed hydrocephalus following intracranial hemorrhage or CNS infection associated with cardiac surgery. These patients were excluded from the study. Eleven children were identified with hydrocephalus and congenital heart disease (table 1). There were 5 males and 6 females. The hydrocephalus was associated with the Dandy Walker malformation (DWM) in 3 cases, aqueductal stenosis in 5 and was idiopathic in 3. Nine children underwent ventriculoperitoneal shunts (VPS), and 1 child had a cystoperitoneal shunt (CPS). In 1 child, the CPS was externalized without complication following the development of ascites from liver and heart failure, and there was 1 shunt infection in another patient.

The cardiac anomalies varied considerably. Five children with relatively simple defects and stable cardiac function were treated medically. Three children with unstable cardiac function required surgery. One child with coarctation of the aorta was diagnosed and treated at 6 years of age.

Of the 11 children, 4 died. One family declined treatment, and the child (K.S.) died at 19 days of age following...
progressive heart failure. An autopsy revealed DWM, polymicrogyria, atrial and ventricular septal defects, cardiomyopathy and 5Q chromosomal deletion. One family declined cardiovascular surgery following a VPS, and the child (N.W.) died of progressive heart failure at 9 days of age. One child with corrected transposition of the great vessels died at 36 months of age from progressive respiratory failure, and 1 child with trisomy 21, DWM and an arteriovenous canal died at 8 months of age following multisystem organ failure. One child (A.P.) is developing normally at 10 years of age. She developed hydrocephalus secondary to aqueductal stenosis at 2 months of age and underwent a VPS; coarctation of the aorta was diagnosed and treated at 6 years of age.

The remaining 6 children have varying degrees of neurodevelopmental delay. Three are severely disabled following hydrocephalus diagnosed within 3 months of age; their cardiac anomalies included patent ductus arteriosus, atrial septal defect and patent foramen ovale. One patient underwent surgical correction. Two patients with idiopathic hydrocephalus showed moderate delay; 1 has a seizure disorder. Both children demonstrated small ventricular septal defects treated medically. One patient (A.K.) shows borderline delay at 2 years of age. This child developed hydrocephalus with a thick cortical mantle and treated with a VPS. His cardiac anomalies were corrected surgically.

Discussion

Analysis of our patients did not indicate a simple relationship between the severity of the congenital heart disease and neurodevelopmental outcome. Clinical series have demonstrated that approximately one quarter of children with congenital heart disease have extracardiac or genetic abnormalities [14, 15], while autopsy reports showed a 10–75% incidence of CNS lesions [16, 17]. Patent ductus arteriosus, complex ventricular wall defects, hypoplastic left heart or pulmonary stenosis were associated with a slightly higher incidence of CNS anomalies. Mortality increases when extracardiac anomalies are present, while children with cyanotic heart disease are at much higher risk for neurodevelopmental delay [8, 9, 11, 14]. Children in our series did not have cyanotic heart disease. One child, patient (A.K.), developed pulmonary hypertension, but the hydrocephalus resulted from a radiologically confirmed aqueductal stenosis and required a VPS. This patient had multiple cardiac anomalies and a developmental examination at 2 years of age indicated borderline delay. This examination may be revised as the patient matures.

However, our series supports previous case reports, describing the poor outcome of children with DWM and correctable cardiac defects [15, 18–21]. The additional risks associated with cardiac surgery have to be evaluated within the context of a potentially poor neurodevelopmental outcome in a child with this congenital malformation [3, 22–25].

Our series also confirms the risks associated with the diagnosis of prenatal hydrocephalus [2, 5, 6, 13, 26, 27]. Two children died within 8 months of age, 1 child showed severe neurodevelopmental delay at 3 years of age, and 1 child showed a moderate delay with a seizure disorder at 4 years of age. These poor results were related to the severity of the hydrocephalus. This risk factor has also been reported in children with idiopathic hydrocephalus or hydrocephalus secondary to aqueductal stenosis [13, 28, 29]. However, the association of complex cardiac anomalies added an additional risk for mortality.

Conclusion

In 7/11 children, the association of symptomatic hydrocephalus and congenital heart disease in the perinatal period resulted in mortality and severe neurodevelopmental delay. In 2/11, this association resulted in moderate neurodevelopmental delay. While the mortality (4/11) of children in this series was equivalent to other series of children with cardiac and extracardiac defects [14, 16, 30], the incidence of neurodevelopmental delay in the survivors (5/7) was higher than in children with isolated hydrocephalus or congenital heart disease. The presence of the DWM or prenatal hydrocephalus indicated a poor prognosis independent of the extent of the cardiac disease. Although an involvement of both organ systems may be infrequent and treated separately, this small series suggested that the burden of congenital disease in this population is substantial and should be taken into account during consideration of treatment options.

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References


