Big heads in Port Moresby General Hospital: an audit of hydrocephalus cases seen from 2003 to 2004

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SUMMARY

Background: Hydrocephalus is a common neurosurgical problem in Port Moresby General Hospital (PMGH) contributing to 27 (24%) of the 114 neurosurgical operations done in 2003 and 2004. During the same period it was responsible for 25% of the cases seen in the neurosurgery clinic. Aim: To prospectively audit and follow up hydrocephalus cases in PMGH over 2 years from January 2003 to December 2004 and ascertain the causes and the outcome of treatment. Method: All cases of hydrocephalus seen in 2003 and 2004 were categorized according to cause. The associated findings on ultrasound scan or CT (computed tomography) scan when available were noted. The subsequent progress was documented with and without treatment for at least 6 months. Results: 61 cases of hydrocephalus were seen for surgical opinion. The age ranged from 4 weeks to 56 years. The commonest age group affected was in the first year of life (61% of cases). There were 34 cases (56%) of congenital hydrocephalus followed by 19 (31%) post meningitis and 8 (13%) due to tumour. There was only one case of myelomeningocele with concomitant hydrocephalus. Venticuloperitoneal (VP) shunts were inserted in 24 cases. 3 shunts were bypasses from the posterior horn to the cisterna magna, making a total of 27 shunt operations. 9 shunts were performed for post-meningitic hydrocephalus, 15 for congenital stenosis and 3 for a posterior fossa tumour. 24 out of the 27 shunt operations were in children aged <9 months. Post-VP-shunt infection of 2 cases reported within 6 weeks of operation gave an infection rate of 7%. There was cerebrospinal fluid (CSF) leak in 2 cases with Pundez-type shunts. There were 2 shunt blocks needing revision. Conclusion: Shunt operations can be done in PMGH with good outcomes. The decision-making about surgery can be made on the basis of the enlarging head and the ultrasound findings.

Introduction

Hydrocephalus is a common neurosurgical problem in Port Moresby General Hospital (PMGH) contributing to 27 (24%) of the 114 neurosurgical operations done in 2003 and 2004 (1,2). During the same period it was responsible for 25% of the cases seen in the neurosurgery clinic (1,2). There is no prior published work on hydrocephalus in Papua New Guinea (PNG).

Aim

The aim of this study was to prospectively diagnose and follow up hydrocephalus cases in PMGH over 2 years in 2003 and 2004 and ascertain the causes and the outcomes of treatment.

Method

All cases of hydrocephalus admitted or seen at the Outpatients Clinic of PMGH from January 2003 to December 2004 were included in the study. These cases were included after clinical assessment of hydrocephalus and confirmation with either ultrasound scan or CT (computed tomography) scan.
Patients were followed up for at least 6 months after surgery or from the first day of consultation to document complications of ventriculoperitoneal (VP) shunts and the outcome by measuring head circumference, neurological progress and serial ultrasound scan assessment of the ventricular sizes.

Results

Distribution of neurosurgery operations in 2 years

Shunt operations contributed to 24% of the neurosurgery operations done at PMGH (Figure 1).

Age and sex distribution

There were 61 cases of hydrocephalus and the ratio of male to female was 31:30. The age range was 1 month to 56 years (Table 1).

Most (75%) of the hydrocephalus cases affected 1-18 month old children, with 61% <1 year old. There were 2 adults – a female aged 56 years with vestibular schwannoma causing hydrocephalus and a male aged 47 with meningioma. The third oldest was a 16-year-old female with aqueduct stenosis. The other cases were <12 years old (Table 1).

Causes of hydrocephalus

The 34 congenital obstructive hydrocephalus cases (Figure 2) were diagnosed by ultrasound scan or CT scan showing dilation only of the lateral and third ventricles and a collapsed or normal fourth ventricle. There was no history of infection or intracranial bleeds and no evidence of mass lesions.

Communicating hydrocephalus (Figure 2) occurred in 19 cases (31%) with previous meningitis, of which 4 cases had active tuberculous (TB) meningitis and were on TB treatment.

8 patients had hydrocephalus due to tumour (Figure 2). These included 1 case of vestibular schwannoma (56-year-old female), 1 case of meningioma (47-year-old male) and 6 children with the following tumours: 2 medulloblastoma, 2 pilocytic astrocytoma, 1 periventricular low-grade glioma and 1 pineal tumour.

Treatment and outcome of hydrocephalus

There were 3 cases of congenital hydrocephalus whose shunts were done earlier in life and required revision due to blockage or shortening distally.

24 cases had ventriculoperitoneal shunts. Out of these, 16 had medium-pressure Hakim valves and 4 had low-pressure valves, two of which were the Pundez burr hole type. The rest were conglomerates of shunt components adjoined into a functioning unit. This is because at one stage we ran out of shunts and had to improvise. 3 shunts were bypasses inserted from the posterior horn to the cisterna magna. This was to treat the hydrocephalus resulting from posterior fossa tumours (Table 2). 24 out of the 27 shunt operations were in children aged less than 9 months.

There were 2 cases of post-VP-shunt infection reported within 6 weeks of the operation (Table 2). No organisms were isolated from our 2 shunt infection cases. The VP shunt was removed in the first case.
TABLE 1

THE AGE AND SEX DISTRIBUTION OF HYDROCEPHALUS CASES AT PORT MORESBY GENERAL HOSPITAL FROM JANUARY 2003 TO DECEMBER 2004

<table>
<thead>
<tr>
<th>Age group</th>
<th>Male</th>
<th>Female</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>0-6 months</td>
<td>13</td>
<td>12</td>
<td>25</td>
</tr>
<tr>
<td>7-12 months</td>
<td>5</td>
<td>7</td>
<td>12</td>
</tr>
<tr>
<td>13-18 months</td>
<td>3</td>
<td>6</td>
<td>9</td>
</tr>
<tr>
<td>19-24 months</td>
<td>2</td>
<td>1</td>
<td>3</td>
</tr>
<tr>
<td>25-30 months</td>
<td>-</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>31-36 months</td>
<td>-</td>
<td>-</td>
<td>0</td>
</tr>
<tr>
<td>3-12 years</td>
<td>7</td>
<td>1</td>
<td>8</td>
</tr>
<tr>
<td>16 years</td>
<td>-</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>47 years</td>
<td>1</td>
<td>-</td>
<td>1</td>
</tr>
<tr>
<td>56 years</td>
<td>-</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>Total</td>
<td>31</td>
<td>30</td>
<td>61</td>
</tr>
</tbody>
</table>

The second case had intraventricular gentamicin for 4 weeks. Both cases improved on antibiotics. The rate of shunt infection was 7% in this series.

Discussion

A VP shunt is indicated in infants where there is increasing ventricular size with cerebral mental thinning less than 2 cm on ultrasound. This is recognized by an occipitofrontal head circumference 2 standard deviations above normal accompanied by further delayed milestones. Urgent shunting is required where there are signs of increased intracranial pressure (ICP) such as hypertonicity or altered consciousness or behaviour. Respiratory compromise and third nerve palsy are late signs. In patients with obvious indications early operation is preferred.

Shunt operations have been done for many years at the PMGH by general surgeons. The outcomes of shunt surgery in PNG have not been previously
TREATMENT AND OUTCOME OF HYDROCEPHALUS IN PORT MORESBY GENERAL HOSPITAL FROM 2003 TO 2004

<table>
<thead>
<tr>
<th></th>
<th>Number</th>
<th>Shunt operations</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Congenital hydrocephalus</td>
<td>34</td>
<td>15</td>
<td>All well</td>
</tr>
<tr>
<td>Post-meningitis hydrocephalus</td>
<td>19</td>
<td>9</td>
<td>2 infections</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>2 leak of CSF*</td>
</tr>
<tr>
<td>Tumour-induced hydrocephalus</td>
<td>8</td>
<td>3</td>
<td>All well</td>
</tr>
<tr>
<td>Total</td>
<td>61</td>
<td>27**</td>
<td>6 complications</td>
</tr>
</tbody>
</table>

*these occurred in Pundez-type shunts
**24 ventriculoperitoneal (VP) shunts and 3 ventriculo-cisterna-magna shunts
CSF = cerebrospinal fluid

Documented. Shunt operations contributed to 24% of the operations done (27 out of 114) in the first two years of a fledgling neurosurgical unit.

The 6 shunt complications were all in cases of malnutrition that had thin skin. In addition the shunts used in 2 of these cases were valves of the burr hole Pundez type with a big dome, prone to skin necrosis, which then leads to CSF leak and infection. Burr-hole-type valves are not recommended in children with thin skin and calvarium.

It is important for health workers to be wary of complications of VP shunts and to report to a surgeon, preferably a neurosurgeon, as soon as possible for any complications.

In the developed world (3) the commonest cause of hydrocephalus is congenital lesions (67%) with or without myelomeningocele. In our series we had 56% (34 cases) of congenital obstruction. The 34 cases had aqueduct stenosis based on the ultrasound findings. This high number of aqueduct stenosis is consistent with the literature, where 70% of cases of congenital hydrocephalus are due to aqueduct stenosis (4). In all the children there was no prior meningitis infection and there were no reports of Chiari malformation. There was no Dandy Walker lesion in this series, which is comparable to elsewhere (3) with a rate of 2.4%.

In this series there was only 1 case of myelomeningocele with hydrocephalus. Elsewhere myelomeningocele is associated with 29% of hydrocephalus (3). The significant difference is probably due to the low frequency of spinal dysraphism in our population, as shown by only 1 case of myelomeningocele in over 20,000 live births in the Labour Ward of PMGH in 2 years (2003 and 2004) (5-8). In high-income countries the rate of myelomeningocele is 20 times greater at 40 per 20,000 live births (9).

There are other birth delivery services outside and within the city of Port Moresby. Accounting for their cases is beyond the scope of this work. It would be interesting to consider a nationwide or regional incidence of spinal dysraphism at some stage. N. Tefuarani, J. Vince, M. Baki and A.B. Amoa (personal communications) affirm the low hospital incidence of spinal dysraphism over the last 25 years despite the high prevalence of folate deficiency in antenatal mothers. The PMGH figures are the best indication for the whole country due to the case-mix of all ethnic groups from all over PNG living in Port Moresby, the national capital. PMGH is the biggest hospital in PNG and has the best ratio of doctors to population. The fact that it is the teaching hospital for all cadres of health workers should enable a better surveillance of conditions such as spinal dysraphism in the newborn. The literature on PNG’s incidence of spinal dysraphism in the past
20 years is sketchy. A prospective study by Dryden of 10,000 consecutive deliveries in PMGH from 1985 to 1986 found only 4 cases of spinal dysraphism (10). A retrospective analysis by Kapanombo from 1987 to 1996 showed 15 cases in 82,515 deliveries at PMGH (11).

The low frequency of spinal dysraphism in PNG is markedly different to that reported from the developed world. This is interesting given the lack of antenatal diagnosis of neural tube defects in general and the non-propagation of therapeutic abortion in PNG; one would have expected a larger caseload of these defects.

Dietary factors such as folic acid have a preventive effect on the prevalence of neural tube defects (12-15). A.B. Amoa and colleagues, from the Obstetrics and Gynaecology Department of the PMGH, reported that 41.5% of antenatal mothers have folate deficiency in one combination or another (16). Despite this low folate intake there is still a low incidence of spinal dysraphism compared to that in children born in high-income countries, who have a folate deficiency rate of 0.2-2% (17) and yet have a higher incidence of spinal dysraphism. Genetics and other environmental factors need to be considered at some stage to explain the low incidence of spinal dysraphism in PMGH.

Meningitis as a cause of hydrocephalus was responsible for 19 (31%) of cases in our series. This is higher than in the developed world with a rate of 7.6% (3) and most probably relates to the incidence of meningitis, and the severity and lack of treatment in babies with meningitis.

The rate of shunt infection was 7% in our series. An acceptable infection rate might be regarded as <10% (18); however, different institutions have their own standards taking into consideration their patient population.

The shunt valve used in our practice is a medium-pressure type in most situations although obstructive hydrocephalus with intra-operatively assessed CSF pressure is best treated with a high-pressure valve. Three cases of shunt operation in tumours included 2 cases of medulloblastoma and 1 case of pilocytic astrocytoma in children. The shunts were inserted via the Frazier burr hole being converted into a bypass from lateral ventricle to cisterna magna when it was realized that the tumours were not fully decompressed due to involvement of the brain stem. Third ventriculostomy is an alternative treatment technique for use in our aqueduct stenosis cases. Although it has great potential the cost of the equipment needed and the steep learning curve required make it impractical at the moment. However, it would save the cost of buying VP shunt components and would prevent shunt infections.

Conclusion and Recommendations

1. Shunt procedures are more likely to be complicated if there is a history of malnutrition and hydrocephalus due to meningitis.

2. Before insertion of a shunt ensure that there are no signs of infection for at least 3 days.

3. Avoid use of a burr hole valve in children. This would best be suited for adults and older children with thicker skull and scalp tissue, which can accommodate the large dome.

4. The low frequency of spinal dysraphism in PMGH needs to be investigated further.

ACKNOWLEDGEMENT

The PNG Tertiary Health Services (THS) Project donated 16 units of Hakim-type valves which are user- and patient-friendly. These shunts have benefited the children of PNG, as we hope this paper has shown.

REFERENCES

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