Pregnancy complicated by maternal hydrocephalus

A report of 3 cases

E. P. FRÖHLICH, J. M. RUSSELL, C. J. VAN GELDEREN

Summary

The obstetric management of 3 women with stable hydrocephalus is presented. The few obstetric complications were unrelated to the maternal hydrocephalus, and specific management of the maternal condition was not necessary. Better management of hydrocephalus in infancy, as well as of the other forms of non-tumoral hydrocephalus, has led to more adults in a stable state with the condition and pregnancy will occur in some of these. A plea is made for all female patients with hydrocephalus to have a ventriculoperitoneal shunt rather than a ventriculostriatal one, with a view to reduction of complications in later pregnancies.

Shunt operations for hydrocephalic patients have been available for more than 30 years. Because of advances in diagnostic procedures and neurosurgical techniques and the availability of unidirectional-flow valve shunts, the prognosis for the non-tumoral hydrocephalus has improved dramatically. An increasing number of these patients, often handicapped with low IQs, different degrees of neurological deficits and even cerebral palsy, reach sexual maturity. Recently, 4 case reports alerted obstetricians about this new group of high-risk patients whose management requires certain genetic and neurosurgical knowledge.

At the Baragwanath Maternity Hospital 3 pregnant hydrocephalic women were admitted between November 1982 and July 1983. The 9 published cases in the English-language literature are reviewed, and our 3 patients added. The pertinent genetic, perinatal, neurological and neurosurgical references are listed. The obstetrician, previously concerned for more than 30 years, because of advances in diagnostic procedures and neurosurgical techniques and the availability of unidirectional-flow valve shunts, the prognosis for the non-tumoral hydrocephalus has improved dramatically. An increasing number of these patients, often handicapped with low IQs, different degrees of neurological deficits and even cerebral palsy, reach sexual maturity. Recently, 4 case reports alerted obstetricians about this new group of high-risk patients whose management requires certain genetic and neurosurgical knowledge.

At the Baragwanath Maternity Hospital 3 pregnant hydrocephalic women were admitted between November 1982 and July 1983. The 9 published cases in the English-language literature are reviewed, and our 3 patients added. The pertinent genetic, perinatal, neurological and neurosurgical references are listed. The obstetrician, previously concerned only with the management of the mother carrying a hydrocephalic fetus, will in future have to be aware of the implications of the mother herself being hydrocephalic.

Case reports

Case 1

A 23-year-old, single, black woman, gravida 2, para 1, was referred to Baragwanath Maternity Hospital in November 1982 for obstetric management. Her first pregnancy in 1978 had been uneventful and had terminated in the vaginal delivery of a 3000 g male infant.

In May 1980, a ventriculoperitoneal (VP) shunt was performed for progressive hydrocephalus caused by a posterior fossa tumour (choroid plexus papilloma). As a result she was blind, with decreased hearing and a mild to moderate degree of motor impairment: clumsiness, poor dexterity, ataxia and left-sided hemiparesis.

The present pregnancy progressed uneventfully to 42 weeks' gestation. The patient was in hospital from 34 weeks' gestation onwards and maternal and fetal well-being were carefully monitored. Consultation with the department of neurosurgery confirmed that the patient's neurological condition was stable so it was decided to terminate the pregnancy. Elective caesarean section was performed under general anaesthesia. No prophylactic antibiotics were used and a healthy male infant weighing 3080 g was delivered through a transverse lower uterine segment incision. The tip of the VP-shunt catheter was seen free in the peritoneal cavity, which was washed out with normal saline. The abdominal cavity was closed in layers. Postoperative recovery was uneventful for both the mother and child. At the 6-week postpartum follow-up visit the mother's condition was stable and the child was thriving.

Case 2

A 15-year-old single black patient, gravida 1, was referred to Baragwanath Maternity Hospital at 38 weeks' gestation. She had had an infantile hydrocephalus treated with a right-sided VP shunt. This had to be replaced in 1977 with a similar device because of a malfunction.

This patient was obviously macrocephalic but of normal intelligence, physically well developed and with minimal neurological deficit: left-sided inco-ordination, dysdiadochokinesis and a left-sided horizontal nystagmus.

An ultrasound examination confirmed the gestational age and excluded major neural tube defects in the fetus. On 29 March 1983 she went into spontaneous labour. A vacuum extraction was performed to shorten the second stage and a live 3280 g female infant was delivered. Mother and infant were well and stable on discharge at 3 days and also at the 6-week postpartum follow-up visit.

Case 3

A 26-year-old single black woman, gravida 1, was transferred to the Baragwanath Maternity Hospital in April 1983. There was inadequate medical information available concerning this patient. She was severely mentally retarded and bedridden because of spastic paraplegia. She was obviously macrocephalic and according to her mother this condition had been evident at birth. A diagnosis of spontaneously arrested hydrocephalus was made.

Results of routine antenatal investigations were all normal and her neurological condition remained unchanged during hospitalization. In July 1983, at an estimated pregnancy duration of 32 weeks, the patient went into premature labour which was successfully arrested with parenteral morphine 15 mg and hydroxyzine 100 mg 6-hourly for 3 doses, and intravenous hexoprenaline 10 μg as a bolus followed by 0,1-0,4 μg/min for 24 hours. Subsequent maintenance therapy was with oral hexoprenaline 0,5 mg sublingual.
usually 6-hourly. No cause was found for the premature labour and despite continued oral hexoprenaline therapy labour recommenced 2 weeks later. Attempted tocolysis failed, and the patient was delivered spontaneously of a 2280 g live female infant. The third stage of labour was complicated by a 1000 ml postpartum haemorrhage controlled by an oxytocin drip; the patient was transfused with 2 U of blood. The remainder of the puerperium was uneventful. The child and mother were stable at the 6-week postpartum follow-up visit.

**Discussion**

Wallman 1 reviewed the history of the management of hydrocephalus with particular reference to the types of shunt employed. He commented that advances in this field may well provide problems for physicians in other fields at a later date. The cases described above are cases in point.

McCullough and Balzer-Martins2 pointed out that approximately two-thirds of treated patients with hydrocephalus will have normal or borderline intellectual capabilities. Jansen et al.3 found that one-third of their patients were leading a normal life although all had signs of neurological impairment. We can conclude that as treatment of hydrocephalus becomes more common and more effective, obstetricians will see such patients achieve maturity — and pregnancy — more frequently.

It then becomes important to consider whether pregnancy, labour (especially where VP shunting has been used) and abdominal surgery have any deleterious effects on the neurological condition.

In all 9 pregnancies in hydrocephalic mothers reported in the English-language literature, and in our 3 cases, the outcome was satisfactory for both mother and infant (Table I). However, there was a high incidence of prematurity (3 out of 12). There were no neurological defects in the 5 infants delivered to women with infantile hydrocephalus 4,7 and our cases 2 and 3.

The management of ventricular shunts requires special precautions.8 One ventriculo-atrial (VA) shunt had to be replaced in early pregnancy owing to malfunction;13 while 2 VP-shunt malfunctions were managed conservatively.11 The overall complication rate as well as severity of the complications is greater in VA shunts than in VP shunts.12

The question of using prophylactic antibiotics when the abdomen has to be opened is as yet unresolved. Howard and Herrick12 recommend their use in all deliveries. In the present series no antibiotics were used and there were no infective complications. However, caution would suggest that there is a place for the use of prophylactic antibiotics when caesarean section has to be performed.

Pregnancy itself requires no special management, but it may be necessary to admit these patients to hospital for prolonged periods in view of their physical and mental handicap. This will depend to a large extent on the patient’s social circumstances.

In view of the high incidence of prematurity it becomes important to know whether labour can safely be inhibited. There were no side-effects from the use of hexoprenaline in 1 of the patients in this series, although the treatment was partially successful.

Management of the second stage of labour was variable, although most authors have recommended assistance. Presumably this has been aimed at reducing intra-abdominal pressure especially in the case of VP shunts. Unidirectional flow valves are an integral part of all modern shunts, and therefore we suggest that possibly this precaution is unnecessary. Certainly, those patients reported by others as well as those in the present series who were allowed to bear down came to no harm. Caesarean section can be performed safely in the presence of a VP shunt but should be employed for obstetrical indications only.

Parenteral, local and regional analgesia in labour have been used (Table I). We are opposed to the use of epidural techniques in the presence of neurological problems. Parenteral analgesia is recommended.

In the majority of cases, neonatal hydrocephalus is a genetic disease4 and in order to counsel these patients, an accurate family history and specific diagnosis on affected infants is essential. Clewell et al.4 first described the surgical approach to the treatment of fetal hydrocephalus and since then there have been sporadic reports of similar aggressive treatment.6,9 The place of such treatment is not yet established10 but it may be that these and similar advances will lead to even more adults with sequelae of congenital disease.

**Conclusions**

Pregnancy, labour and delivery are safe in patients with successfully treated hydrocephalus. The only special treatment required will relate to each patient’s specific mental, physical and social circumstances. Complications are rare and the outcome is generally good. We can anticipate an increase in the numbers of such patients in the near future. We

---

**TABLE I. SUMMARY OF REPORTED CASES**

<table>
<thead>
<tr>
<th>Series</th>
<th>No. of patients</th>
<th>Type of shunt</th>
<th>Complications</th>
<th>Prophylactic antibiotics</th>
<th>Mode of delivery</th>
<th>Outcome</th>
<th>Maternal</th>
<th>Fetal</th>
</tr>
</thead>
<tbody>
<tr>
<td>Monfared et al.14</td>
<td>1</td>
<td>VA</td>
<td>None</td>
<td>Parenteral</td>
<td>Forceps</td>
<td>No</td>
<td>Good</td>
<td>Good</td>
</tr>
<tr>
<td>Howard and Herrick12</td>
<td>1</td>
<td>VP</td>
<td>None</td>
<td>Parenteral</td>
<td>Forceps</td>
<td>No</td>
<td>Good</td>
<td>Good</td>
</tr>
<tr>
<td>Kleinman et al.11</td>
<td>1</td>
<td>VP</td>
<td>None</td>
<td>Parenteral</td>
<td>Forceps</td>
<td>No</td>
<td>Good</td>
<td>2380</td>
</tr>
<tr>
<td>Gast et al.13</td>
<td>1</td>
<td>VA</td>
<td>None</td>
<td>Regional</td>
<td>Forceps</td>
<td>Yes</td>
<td>Good</td>
<td>Good</td>
</tr>
<tr>
<td>This series</td>
<td>1</td>
<td>VP</td>
<td>None</td>
<td>Parenteral</td>
<td>Vacuum</td>
<td>No</td>
<td>Good</td>
<td>Good</td>
</tr>
<tr>
<td></td>
<td>1</td>
<td>VA</td>
<td>Replaced</td>
<td>Local</td>
<td>Vaginal</td>
<td>Yes</td>
<td>Good</td>
<td>Good</td>
</tr>
<tr>
<td></td>
<td>1</td>
<td>VP</td>
<td>None</td>
<td>Parenteral</td>
<td>C/S</td>
<td>No</td>
<td>Good</td>
<td>Good</td>
</tr>
<tr>
<td></td>
<td>1</td>
<td>VP</td>
<td>None</td>
<td>Parenteral</td>
<td>Vaginal</td>
<td>No</td>
<td>Good</td>
<td>2280</td>
</tr>
</tbody>
</table>

VA = ventriculo-atrial shunt; VP = ventriculoperitoneal shunt; Prem. = premature birth; GA = general anaesthesia; C/S = caesarean section.
would recommend the use of VP shunting in female patients whenever possible.

We should like to thank Mrs V. Steyn for typing the manuscript and Mr P. Atwell for compiling the table.

REFERENCES


Graves' disease and selective resistance of pituitary thyrotroph to thyroid hormone

A case report

N. L. PILLAY, I. JIALAL, P. A. GOVENDER, S. M. JOUBERT

Summary

A patient with Graves' disease who had inappropriately increased thyroid stimulating hormone (TSH) concentrations and TSH response to thyrotrophin-releasing hormone is reported. No pituitary tumour could be demonstrated and the patient is considered to be an example of selective resistance of the pituitary thyrotroph to thyroid hormone action.

The response of thyroid-stimulating hormone (TSH) to intravenously administered thyrotropin-releasing hormone (TRH) is regarded as a sensitive test in the definitive diagnosis of primary hyperthyroidism: in patients with primary hyperthyroidism and exophthalmic Graves' disease the response is absent or impaired, while a normal response excludes the diagnosis of hyperthyroidism. However, in some cases of hyperthyroidism associated with inappropriate TSH secretion, the TSH responses can be normal. A patient with all the features of Graves' disease, who manifested inappropriately high TSH secretion and showed a brisk TSH response to intravenous TRH, is reported.

Case report

The patient, a 24-year-old black woman, 24 weeks' pregnant, presented at the antenatal clinic, King Edward VIII Hospital, with clinical features of hyperthyroidism. Before her pregnancy she had lost weight (in spite of increased appetite), sweated excessively, preferred cold weather, had palpitations and was restless and anxious. Eight years previously she had been treated for hyperthyroidism but had defaulted after a few months, and 3 years previously she had had a spontaneous abortion at 20 weeks' gestation. The patient's younger brother, aged 12 years, had also been treated for hyperthyroidism.

Examination revealed an asthenic woman with exophthalmos (no ophthalmoplegia), marked restlessness and hyperactivity. She

MRC Preclinical Diagnostic Chemistry Research Unit, Department of Chemical Pathology, and Department of Obstetrics and Gynaecology, University of Natal and King Edward VIII Hospital, Durban

N. J. PILLAY, M.B. CH.B., F.F.PATH.
I. JIALAL, M.D.
S. M. JOUBERT, M.B. CH.B., M.R.C. PATH.