OPHTHALMIC COMPLICATIONS OF MENINGOMYELOCELE AND HYDROCEPHALUS IN CHILDREN*†

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MENINGOMYELOCELE is a major developmental malformation which occurs in about 2·5 per thousand live births in Great Britain (Nash, 1963). A cystic swelling formed by dura and arachnoid mater protrudes through a defect in the vertebral arches (Fig. 1) and this herniated sac contains spinal cord tissue or spinal nerve roots which are attached to the fundus of the sac (Cameron, 1956). As a result there is usually a marked loss of motor and sensory function below the level of the lesion (Sharrard, 1963). 82 per cent. of meningocele are lumbar or sacral in site (Eckstein and Macnab, 1966) so that there is paralysis and anaesthesia of the lower limbs. Defective sensation of the bladder and rectum is accompanied by incontinence and, in the case of the urinary tract, retention and back pressure leading to progressive renal deterioration (Nash, 1956). Because of an associated Arnold-Chiari malformation, 83 per cent. of cases show ventriculographic evidence of hydrocephalus in the perinatal period (Lorber, 1961), and in more than half these cases the hydrocephalus is rapidly progressive in the first 3 months of life.

FIG. 1.—Transverse section through meningocele sac showing defect in a vertebral arch and spinal cord tissue exposed on the body surface.

Until recently the outlook for life in these children has been extremely poor because, although in the majority of cases the meningeal defect could be closed, the progressive hydrocephalus was unlikely to be spontaneously arrested so that surviving infants were mentally retarded largely through cerebral cortical thinning. Such a gloomy prognosis has now been radically altered by the development of the Spitz-Holter and Pudenz-Heyer valves, permanent one-directional Silastic reed-check valves with a constant opening

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FIG. 2.—Valve and catheter used to establish a ventriculo-atrial shunt.

FIG. 3.—Right ventriculo-atrial shunt in position.

pressure, usually 10 mm. of water (Fig. 2). A catheter is introduced into the lateral ventricle and attached to the inlet side of the valve, while a second catheter connects the outlet side to the right atrium of the heart (Fig. 3). In this way, controlled drainage of cerebrospinal fluid is established with little danger of retrograde infection (Macnab, 1962). Affected children are now transferred to special surgical centres where the spinal defect is repaired on the day of birth (Nash, 1963), so reducing the risk of meningitis and improving the function of the lower limb musculature (Sharrard, Zachary, Lorber, and Bruce, 1963). If, during the following few weeks, an excessive increase in skull circumference is observed, the hydrocephalus is brought under control by the insertion of a ventriculo-atrial shunt. These procedures have greatly increased the chances of affected children surviving beyond infancy (Eckstein and Macnab, 1966), and as a result the secondary problems of urinary retention and incontinence, recurrent urinary tract infection, trophic ulceration, and limb deformity resulting from the paraplegic state now present the main challenge. The rewards of expert and energetic treatment are great for, although many of these afflicted children require special residential care and education, the majority are of normal intelligence (Stephen, 1963). During the past 6 years the regime described above has been followed in the treatment of more than seventy affected children admitted to St. Bartholomew's Hospital, London.

Both in patients who have had ventriculo-atrial drainage procedures and in those in whom there was no progressive developmental hydrocephalus there is a long-term risk of two serious intracranial complications: infection and a secondary rise in intracranial

pressure. Meningitis and encephalitis may occur by direct infection through the site of the meningocele. A secondary rise in intracranial pressure may follow blockage of the artificial drainage system at its upper or lower end, while meningitis may also give rise to interference with the normal circulation of cerebrospinal fluid and so precipitate secondary hydrocephalus. In addition, chronic urinary tract infection may cause meningitis by blood-borne spread, and a secondary rise in intracranial pressure by precipitating either a toxicephalopathic state or toxic cardiopathy causing right-sided cardiac failure and consequent back pressure on the ventriculo-atrial drainage system. Any rise in intracranial pressure will be most severe if there has been previous premature fusion of skull sutures due to a period of intracranial hypotension following the original ventriculo-atrial drainage procedure, as suggested by radiographic appearances of partial synostosis associated with springing of other cranial sutures (Fig. 4). Delayed hydrocephalus may be either acute or insidious in onset; in the former case the symptoms of malaise, vomiting, and fever may mimic those of severe urinary tract infection, of meningitis, or of any febrile illness. In the latter, there may be little general disturbance.

FIG. 4.—Lateral x ray of skull of a 2-year-old child with a blocked ventriculo-atrial shunt and secondary rise in intracranial pressure, showing partial synostosis of the fronto-parietal suture, springing of the occipito-parietal suture, and digital markings.

Ophthalmic Complications

This introductory survey indicates that affected children run the risk of a number of ophthalmic complications. Meningitis or encephalitis may damage the optic nerves, the higher visual pathways, or the visual cortex, giving rise to primary optic atrophy or cortical blindness, and may precipitate strabismus from extrinsic ocular muscle palsies. A rise in intracranial pressure may cause papilloedema leading to secondary optic atrophy, or a dilatation of the third ventricle, or a shift in the position of the brainstem giving rise to a primary optic atrophy from stretching of the optic nerves or optic chiasma. Extrinsic ocular muscle palsies may also occur as a non-localizing sign of raised intracranial pressure. It should be stressed that, although the majority of affected children do not suffer from visual complications, these nevertheless remain a threat which may add a further dimension to the child's already severe degree of physical handicap.

Strabismus is a common finding in these children, affecting 30 per cent. of cases in this series. Its presentations will be analysed more fully in a subsequent communication, but briefly it is most commonly an incomitant convergent squint of the alternating type, due to sixth nerve paresis as a result of the abnormal ante-natal cerebral development, and it may be regarded as a "congenital" form of squint as it is present so early in life. Less commonly a sixth nerve palsy may follow meningitis or a secondary rise in intracranial pressure with the development of a rapidly increasing convergent squint; indeed the squint may provide a good non-localizing sign of the intracranial hypertension. A concomitant squint, usually of the convergent type, may arise either as a result of defective vision or, in mentally retarded children with poor fusion faculty, at a later stage.

Case Reports

Case 1.—This child was born 5 weeks prematurely in January, 1962. He had a large lumbo-sacral meningocele (Fig. 5) and was transferred to St. Bartholomew's Hospital 6 hours after birth where the spinal defect was repaired on the same day (Fig. 6). At that time the skull circumference was recorded as 32 cm., but 3 weeks later it was evident that hydrocephalus was developing, the skull circumference having increased to 37 cm. A Spitz-Holter valve was inserted in a shunt between the left lateral ventricle and the pleural cavity. After this operation the hydrocephalus was well controlled, and when the child was 5 months old the lower end of the shunt was transferred to the right atrium *via* the left internal jugular vein. The child remained well and in July, 1965, when he was 3½ years old, an orthopaedic operation of soft tissue release of the left hip and bilateral elongation of the Achilles tendon was carried out in order to correct the contraction deformity of the lower limbs caused by the paraplegic state. A month later the child developed an acute illness characterized by fever, malaise, and vomiting. His valve was found to be emptying only slowly and a course of systemic antibiotic therapy was instituted. Ophthalmic examination revealed an alternating convergent squint with bilateral limitation of abduction. There were normal light fixation and following reflexes, normal pupillary light reflexes, and no fundus abnormality was noted. During the following few days roving eye movements developed, the pupillary light reflexes became sluggish, and bilateral papilloedema developed. An emergency revision of the

FIG. 5.—Large lumbo-sacral meningocele before operation (Casl).

FIG. 6.—The same patient after operation.

ventriculo-atrial shunt was undertaken, but the papilloedema did not resolve and 2 weeks later the valve was again explored. After this second operation the oedema subsided, but secondary optic atrophy developed, and since that time vision has been limited to vague perception of light in both eyes.

Case 2.—This child's early history is similar to that of the first case. She was born in February, 1963, with a thoraco-lumbar meningocele which was repaired on the first day of life, and a Spitz-Holter shunt was subsequently inserted at the age of 3 weeks. An alternating convergent squint with bilateral limitation of abduction was first noted in August, 1966, when she was 3 years old, and at that time ophthalmic examination showed normal fixation and following reflexes in both eyes, normal pupillary light reflexes, and healthy fundi. Bilateral medial rectus recession operations were carried out. A month later she developed an acute febrile illness with marked irritability and some neck stiffness. At another hospital a lumbar puncture revealed sterile cerebrospinal fluid at normal pressure with a raised protein content and increased lymphocyte count. The Spitz-Holter valve appeared to be functioning correctly, and visual function and fundus appearances were also reported as normal. A diagnosis of viral meningitis was made and the child's condition improved at first, but relapsed a week later when the intracranial pressure was thought to be increased and for the first time there was evidence of deterioration in vision. The patient was transferred to St. Bartholomew's Hospital where her lack of response to visual stimuli was confirmed; there were poor light fixation and following responses, and the pupillary light reflexes were sluggish, but the optic discs were normal in appearance. The Spitz-Holter shunt was explored and a blocked catheter was replaced. The child's vision did not improve and 4 weeks after the onset of visual symptoms signs of primary optic atrophy first became visible ophthalmoscopically. The visual status has remained unchanged since that time; there is perception of hand movements only with each eye, and marked pallor of both optic discs.

Case 3.—This child was born in May, 1964, with a dorso-lumbar meningocele which was repaired on the first day of life. A Spitz-Holter shunt was inserted when the child was 3 weeks old. The drainage mechanism had to be revised on a number of occasions and he also suffered from several episodes of urinary tract infection. At the age of 2 years he was referred to the Eye Department because of an alternating convergent squint. There was a full range of ocular movements, and no abnormal fundus appearances were noted. A left medial rectus recession and lateral rectus resection were carried out. A month after this operation the patient suffered an acute febrile illness with malaise, vomiting, and the appearance of a rash. The diagnosis of measles was made, but at the same time it was noted that the Spitz-Holter shunt was not filling well, and ophthalmoscopic examination revealed the presence of a moderate degree of bilateral papilloedema. Because of the patient's poor general condition revision of the drainage mechanism was not attempted at this stage, but the meningocele sac was tapped on a number of occasions. During the following month the papilloedema slowly subsided without the supervision of secondary optic atrophy. There was no evidence of impaired visual function at any time and subsequently, when the child's general condition improved, the drainage mechanism was successfully revised.

Case 4.—This child was born in April, 1965, with a low thoraco-lumbar meningocele which was repaired on the day of birth. A ventriculo-atrial shunt was successfully carried out when he was 3 weeks old because of progressive hydrocephalus. At the age of 2 years he suffered an acute febrile illness with nausea and drowsiness but no neck stiffness or photophobia. Ophthalmoscopic examination revealed no evidence of papilloedema, but clinically the Spitz-Holter valve appeared to be blocked and revision was carried out. At operation the pressure in the lateral ventricle was found to be 200 mm. cerebrospinal fluid. After this operation the patient's general condition improved and at no time has there been any evidence of impaired visual function.

Discussion

The first two cases described demonstrate the difficulty of differentiating between the symptoms of meningitis and those of rising intracranial pressure, and it is likely that in both cases the two conditions co-existed. In the first case papilloedema was the first unequivocal indication of raised intracranial pressure and it was tragic that despite emergency cranial decompression secondary optic atrophy developed. In the second case there is some doubt as to the exact cause of the primary optic atrophy, but it seems likely that a secondary rise in intracranial pressure was precipitated by meningitis and that this possibly caused stretching of the optic chiasma. The fact that atrophic changes were late to appear at the optic nerve head after the onset of visual symptoms certainly suggests that the initial damage occurred fairly far back along the course of the optic nerve fibres. In the third case, there was again a clinically confusing situation, further complicated by an intercurrent exanthematous infection. Ophthalmoscopic examination revealed the presence of papilloedema at an early stage before secondary atrophic changes in the nerve head had occurred and this finding determined an emergency cerebrospinal decompression even although the child's general condition prevented any extensive surgical procedure. The fourth case emphasizes the point that the appearance of papilloedema, indicating a rise in intracranial pressure, makes surgical intervention a matter of urgency to avoid the risk of secondary optic atrophy, but the absence of papilloedema does not necessarily mean that the pressure is normal and should not lull the surgeon into a false sense of security. Of great interest is the fact that in three of these four cases the secondary rise in intracranial pressure occurred soon after an operation involving a general anaesthetic. It may be that during such a procedure there is a risk of displacement or derangement of the valve itself, or of distortion or fracture of the ventricular catheter. It follows that not only should great gentleness and care be used in the positioning of the head during such operative procedures, but that the patient should be carefully observed during the post-operative period for the possible appearance of early symptoms and signs of raised intracranial pressure.

Conclusion

The ophthalmologist has an important role to play in assisting the paediatric surgeon in the long-term management of children with meningomyeloceles if the risk of blindness is not to be added to their other physical disabilities. These patients differ from those suffering from the effects of developmental hydrocephalus only, which has been arrested by a ventriculo-atrial drainage procedure, and pose a much more complex clinical problem. The severe urinary tract infections to which they are susceptible may mimic and in some cases mask the symptoms of rapidly rising intracranial pressure. The finding of papilloedema or, more commonly, of a rapidly increasing angle of incomitant convergent squint may then be the first unequivocal signs of the latter complication. When a secondary rise in intracranial pressure occurs slowly and insidiously there may be no systemic symptoms, so that the finding of low grade papilloedema is the first definite sign of this complication. In addition, if a child's general condition is poor because of intercurrent urinary or other infection, conservative management of an episode of raised intracranial pressure may have to be considered provided there appears to be no immediate risk to visual function, in particular from the development of secondary optic atrophy. For all these reasons it is clear that frequent examinations of the eyes ought to be established as an important part of the long-term management of these children, special attention being paid to the range of

extrinsic ocular movements, the presence of squint, the appearance of the optic discs and, if possible, the extent of the fields of vision. Finally, strabismus surgery plays an important part in the rehabilitation of many of these children. Even if binocular function is unlikely to develop, the cosmetic correction of a marked squint is of great importance both to the young patient and to his parents because it removes one obvious aspect of the child's deformity.

Summary

The modern management of meningocele, which is commonly associated with developmental hydrocephalus, is described. The ophthalmic complications which may occur as a result of this condition are discussed and four cases are described which illustrate the mechanisms by which these complications may arise. The importance of regular ophthalmic examination in the long-term management of these children is stressed.

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