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Case reports

Management of pregnancy in mothers with spina bifida

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Summary

Pregnancy complicated because of maternal spina bifida remains uncommon. With more aggressive surgical management in the 1960s these patients are now in adulthood and achieving pregnancies. In this series all four patients were delivered by caesarean section. As most patients have undergone extensive urological surgery great care is required at operation to avoid trauma to the urinary tract.

Maternal spina bifida; Pregnancy

Introduction

Improvements in corrective surgery in infancy for spina bifida cystica and its related complications have allowed more such women to achieve a pregnancy on reaching adulthood. This has certainly been our local experience and we have had opportunity recently to contribute to the management of four such women.

Problems in pregnancy described in the literature would appear to be related to urinary tract complication; in particular, infection and problems of accommodation of the growing uterus within the abdomen in the presence of a restrictive kyphoscoliosis [1–3].

We report the problems encountered in four women with similar basic spinal lesions delivered within a period of 12 months in the Liverpool city hospitals.

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Case 1

Mrs. H.M. is 28 years old and was born with a lumbar meningocele. Complications and surgical procedures in infancy included closure of the lesion, removal of a lumbosacral teratoma, below-knee amputation due to trophic ulceration, and a neuropathic bladder which necessitated ileostomy and ureteric diversion. The latter failed after ten years and had to be resited. She was left with bilateral hydronephrosis and hydroureter although with only minimal compromise of renal function.

In 1984 she had had a 12 week spontaneous abortion, followed the same year by irrevocable preterm labour at 26 weeks gestation. This resulted in the vaginal delivery of a baby girl who died from hyaline membrane disease soon after delivery. In the present pregnancy she booked at 12 weeks and at 16 weeks her serum alphafetoprotein was normal.

Urinary tract complications

Despite appropriate therapeutic and then prophylactic antibiotic therapy she experienced four recurrent infections of *E. coli* organism during the pregnancy and two recurrent infections with *Pseudomonas aeruginosa*. She was admitted at 26 weeks gestation in threatened preterm labour in association with an exacerbation of one of the urinary tract infections. This responded to intravenous ritodrine. She was also found to have a hypochromic, microcytic anaemia with normal serum iron, folate and B12.

Clinical pelvimetry revealed minimal restriction of the pelvic diameters, and a broad plan for vaginal delivery was made. Prior to this a consultation had been made with her urological surgeons to ascertain the nature of the refashioned ileal loop diversion and its relation to the uterus. However, a persistent breech presentation was noticed and when spontaneous preterm labour commenced at 33 weeks with spontaneous rupture of the membranes it was thought wisest to perform a caesarean section. At caesarean section the lower segment was reasonably formed and a baby girl was delivered with ease, weighing 2500 g. The Apgar scores were 2 at 1 min and 7 at 5 min. She progressed well in special care.

A mid-line incision had been made in case of adhesions following previous bladder and ureteric surgery. At the end of the caesarean section, inspection of the ileal loop revealed that it had been inadvertently damaged during the peritoneal incision, and a consultant urologist colleague was summoned to assist with the repair.

The loop was catheter-drained for the following ten days and her recovery was uneventful save for a further *E. coli* urinary tract infection.

Case 2

Mrs. H.B. This lady was a 28-yr-old primigravida who was born with a myelomeningocele which was closed at birth. In childhood she required several foot operations which preserved her ability to walk although with appreciable difficulty. Urological investigations in late childhood revealed a non-functioning right kidney with compensatory hypertrophy of the left kidney. She also had a hypotonic bladder

and seemed to control micturition more by a process of overflow incontinence than genuine voiding.

The pregnancy was complicated by one urinary tract infection. The pelvic measurements which were assessed clinically and radiologically were reduced and plans were made for elective caesarean section.

Spontaneous labour, however, occurred at 37 weeks and a short trial of labour was unsuccessful, culminating in routine lower-segment emergency caesarean section. She was delivered of a baby boy weighing 2.49 kg who was well at birth and continued to thrive.

In the puerperium she had two episodes of urinary retention in association with *E. coli* infection. She responded to bladder drill and pyridostigmine but still was left with a hypotonic bladder, once more seemingly controlling her micturition by a form of overflow incontinence. She was referred for further urodynamic studies with a view to clean, intermittent self-catheterization.

Case 3

Mrs. C.B. This 29-yr-old primigravida had primary closure of her myelomeningocele and several orthopaedic operations in childhood. She suffered from an uncorrected neuropathic bladder and an element of kyphoscoliosis.

The antenatal period was relatively uneventful though the neuropathic nature of her bladder function proved increasingly distressing. She was admitted for rest and assessment at 29 weeks gestation. Digital and radiological pelvimetry measurements were only slightly reduced, but the presence of the kyphoscoliosis favoured a persistent parietal presentation.

Elective caesarean section was performed at 38 weeks, resulting in the birth of a baby boy weighing 3.55 kg with Apgar scores of 9 and 10 and 1 and 5 min respectively. She made an uneventful recovery and was discharged.

Case 4

Mrs. J.M. This 19-yr-old patient was in fact born at the same hospital in 1966 with myelomeningocele and hydrocephalus. The myelomeningocele was closed soon after birth and a Spitz-Holter valve was inserted to drain the hydrocephaly. A bilateral ureterostomy was performed in early childhood to preserve renal function, which was deteriorating as a result of ureteric reflux from her neuropathic bladder and persistent urinary tract infections. The stoma was sited in the midline midway between the umbilicus and symphysis pubis. She subsequently required a cystectomy for persistent infective cystitis. Mrs. J.M. underwent numerous orthopaedic procedures and managed to walk with difficulty until the age of fifteen using crutches, having then to resort to a wheelchair.

This pregnancy was complicated by recurrent *Proteus* urinary tract infections and she was treated with long-term penicillin. Her renal function was monitored, with regular estimations of urea and electrolytes and also creatinine clearance. The latter varied from 69 ml/min to 122 ml/min. Her general health was satisfactory throughout the pregnancy and she remained normotensive. She did not wish

alphafetoprotein measurements to be made. Clinical pelvimetry revealed a small pelvic inlet and it was decided to perform elective caesarean section at 38 weeks gestation. A ureterogram was performed the day before operation to outline the ureters, which lay extraperitoneally. At operation two 8 French gauge catheters were inserted, one along each ureter, to outline their position. A low transverse incision was performed and Mrs. J.M. was successfully delivered of a live male infant weighing 3.18 kg with Apgar scores of 9 and 10 at 1 and 5 min. She made an uncomplicated recovery and was discharged from hospital with an appointment for urological review at 6 weeks.

Discussion

Sporadic cases of pregnancy occurring in patients with congenital spina bifida have been reported [1–10]. Carter and Evans [4] assessed the risk to offspring of parents of either sex with spina bifida cystica at 3%. They concentrated primarily on the genetic inheritance of this condition and made no reference to maternal antenatal complications or modes of delivery. Laurence and Berisford [6] estimated the incidence of a neural tube defect in the offspring of either parent suffering from the condition as 1 in 23, although the risk of having an affected offspring differed for sons and daughters (1 in 50 and 1 in 13 respectively). This paper described 56 cases of adult spina bifida but concentrated on the social aspects of marriage and continence. Minimal attention was paid to pregnancy and the mode of delivery.

Most reports concentrate on single case presentations. However, Wynn et al. [1] presented three cases with satisfactory outcomes, all ending in vaginal delivery. Although caesarean sections should only be considered for purely obstetric reasons, this is often the preferred mode of delivery [3]. A single case of a vaginal delivery under epidural block has been reported [11] but this appears to be the exception rather than the rule.

The main risks are those of urinary tract infection, accommodation problems for the fetus which may result in mechanical difficulties with delivery, an increased risk of a neural tube defect in the offspring and the social problems related to coping with a new-born infant.

Nowadays, myelomeningocele and paraparesis are not contra-indications for pregnancy. If possible, these patients should be counselled prior to conception, for assessment of renal function and radiological assessment of pelvic dimensions could be made without risk to the fetus in-utero. Renal function should obviously be monitored closely throughout pregnancy with regular bacteriological assessment of the urine. If corrective surgery has been required to treat urinary incontinence, which is nearly always due to a neurogenic bladder, then we would recommend some form of ureterogram prior to delivery, particularly if caesarean section is planned.

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