

Gastroschisis in Northern Australia. Is the experience transferable?

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Introduction

Gastroschisis is a congenital abnormality in which there is a full thickness defect in the anterior abdominal wall through which intestines and sometimes other organs protrude into the amniotic sac before birth. The defect is usually of small size, to the right of the umbilicus from which it is separated by normal skin, and unrestricted by any covering membrane¹, as compared to herniation through the umbilicus which is known as omphalocele. The incidence is increasing at such a rate in certain countries the appellation 'epidemic' has been applied though no-one has found an underlying cause for the apparent failure of closure of lateral body wall folds or of the yolk sac to be incorporated into the umbilical stalk.

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Epidemiological trend

Various associations have been noted: especially younger maternal age and lower socio-economic status, but more weakly, lower maternal BMI, exposure to vaso-active drugs including tobacco, multiple sexual partners and peri-conceptual infection².

No racial predominance had been detected until review of the incidence in Australian Indigenous mothers revealed it to be twice as high as in non-Indigenous Australians in the rural region of North Queensland where many Indigenous families live in reduced circumstances³. When considered together over a twenty year period, the incidence has risen from 0.7 to almost 4.8 per 10,000 live births. In Indigenous mothers under 20 years of age, however, it rose to almost 19.3 per 10,000: the highest reported rate in any Indigenous group in the world. This rate is likely, in fact, be higher because it does not include stillbirths or miscarriages.

Problem statement

It is not known why gastroschisis should be so high in young Indigenous Australians, though most of the reported associations are relevant. The magnitude of peri-conceptual infections in this population suggests a vascular effect of cytokines on the arteries supplying the anterior body wall but why this effect should be confined to those arteries is, of course, unknown and not well understood.

Case finding

Gastroschisis is traditionally considered a single defect compared with omphalocele and its likelihood of multiple problems. Recent studies have found an unexpected association of gastroschisis with other defects⁴. Though the association of undescended testicle might be explained as secondary to reduced intra-abdominal pressure, other defects including heart and limb defects are less explicable. It is evidenced that 14% of cases in North Queensland have in addition other abnormalities.

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Fig. 1 Showing Protrusion of intestine through anterior abdominal wall in a gastroschisis of a new born

Experimental study

Treatment

Though little new has emerged with regard to the demography (apart from its rising incidence) and underlying cause of gastroschisis, the treatment has altered radically, and for the better, in North Queensland. Though the morbidity and mortality rates have not changed, the ease of operation has been transformed by the simple technique of manually replacing the herniated bowel soon after birth and closing the defect by pulling the umbilical cord across it and applying a transparent, adherent dressing such as Op Site to hold it in place. The dressing is applied until the defect heals within a week or two. In the meantime the baby is fed intravenously⁵. This technique has been described by Sandler et al⁶.

We reviewed our management of 50 cases of gastroschisis which had occurred over a twenty year period. 16 cases were treated by this primary non-operative care and 22 were treated by primary operative care comprising formal resection of the defect, replacement of the herniated bowel and suturing the defect in fascial and skin layers in the theatre. We found no significant difference in associated length of stay (about 4 weeks), duration of intravenous feeding (about 3 weeks) or other medical and surgical complications or outcomes. Mode of delivery (normal vaginal versus Caesarean) made no difference to outcomes.

All of these 38 cases were defined as non-complex. Complex cases included those in which the bowel could not be replaced without significant fear of vascular compromise because of the increased pressure within the abdominal cavity. There were 6 of these cases and they were treated by fitting a protective silo (originally constructed by a 'bile bag' and then supplied by Bentec Medical, Woodland, US) to allow staged reduction of the bowel over a period of about a week when the defect was closed by suturing fascial and skin layers in the theatre. More complex cases included those with significant matting, twisting or perforation or atresia of the bowel, and these were managed by appropriate operative repair of the bowel (with or without attendant ileostomy) and closure of the defect in the theatre.

Three babies died giving a mortality rate of 6%. None died after primary non-operative care. One died from associated pulmonary hypoplasia and two after complications with extensive areas of atresia.

The advantage of the primary non operative care was that it could be performed in the nursery without taking the child to theatre. It could thus be performed quickly with minimal heat loss, exposure to the risk of infection, and less vascular compromise to a bowel suspended outside the abdominal wall while awaiting operation. The child could be kept warm, hydrated intravenously and sedated with morphine to minimise abdominal wall tension while the bowel was replaced without formal anaesthetic.

Management

Once the bowel is replaced, management depends on provision of nutrition by intravenous means, and careful nursing to maintain warmth and detection of infection until feeds are established. Adjustment of the volumes of parenteral nutrition prepared for adults offers the possibility of nutrition, while minimising the cost of local preparation and the attendant risk of infection. The cost and difficulty of parenteral nutrition for several weeks is, however, not underestimated. This provision of nutrition would appear to be the

greatest obstacle to successful management of non-complex gastroschisis in countries with greater burdens of disease.

Conclusion

In many parts of the world, the use of silos is the preferred form of management for all gastroschisis, including the non-complex cases, because of the worry of compartment syndrome. We found no added risk of this complication. Silos, moreover, are expensive and demand close nursing attention. We believe non-operative primary replacement is a simple procedure that is appropriate for almost three quarters of the cases. Those for which it is not appropriate are usually obvious, involving matted, perforated, twisted bowel. Therefore, it appears a reasonable approach for hospitals where resources are challenged. We want to see that our experience be applied elsewhere.

Reference

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