



Best practice guidelines

Abdominal wall defects



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ABSTRACT

Abdominal wall defects in fetuses include gastroschisis, exomphalos, bladder exstrophy complex, cloacal exstrophy and body stalk syndrome. The defects that occur more commonly are gastroschisis and exomphalos. In this review we assess the current evidence regarding the incidence, perinatal risk factors, antenatal and post-natal management and outcome for both these conditions. A review of the current surgical practices for management of gastroschisis and exomphalos is discussed.

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1. Introduction

Until early 1940, all anterior abdominal wall defects were classified under gastroschisis and there was no clear understanding of the underlying pathology or management of these defects. Invariably, the mortality at that time was close to 90–100%. Since then further efforts at classification of nomenclature and management of these defects were made. The fundamental differences in the conditions and the risk factor were identified. Over the decades, the incidence of gastroschisis has

been increasing. With improvement in antenatal screening for foetal anomaly, most cases are now identified antenatally and managed by an expert multidisciplinary team.

This review article aims to identify the current epidemiology, perinatal management and surgical management and outcome of common anterior abdominal wall defects, mainly gastroschisis and exomphalos.

2. Gastroschisis

2.1. Epidemiology – incidence and risk factors

The incidence of gastroschisis has increased since 1995 is 4.42 per 10,000 live births [1]. Maternal age below 20 years is a major risk factor

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for gastroschisis with an incidence of 11.45 per 10,000 live births. The other risk factors for gastroschisis are primigravida status, low socioeconomic status, change in paternity, poor nutrition, smoking and substance abuse by either of the parents. Maternal obesity may be protective [2].

2.2. Diagnosis

Foetal ultrasound is highly sensitive in identifying anterior abdominal wall defects. Contemporary techniques can predict complicated gastroschisis and outcome measures that can be correlated with degree of antenatal dilatation. Intra-abdominal bowel dilatation >14 mm is predictive of bowel atresia in 29% of cases [3]. The use of magnetic resonance imaging is limited in gastroschisis and in certain cases can be used as an adjunct to evaluate bowel atresia in gastroschisis [4].

2.3. Antenatal management

There is inadequate evidence about optimal antenatal surveillance and current practice is variable. There is some evidence that ultrasound monitoring of growth, umbilical artery Doppler and bowel diameter measurements lead to early detection of complications and can help improve mortality [5]. However, a recent systematic review looking at the outcome of antenatal bowel dilatation showed no difference in mortality, length of bowel resection, time to feeding or length of hospital stay [6].

2.4. Outcome

2.4.1. Delivery method

Labour is routinely induced at 37 weeks gestation or earlier if foetal well-being is abnormal. In a single perinatal centre, comparing induced and expectant labour, 20% proceeded to caesarean section in both groups [7]. Although, there was decreased incidence of respiratory distress syndrome and improved Apgar scores in the expectant group, the incidence of sepsis doubled and bowel damage was tripled compared to induced labour. With induced labour there was the added advantage of shorter time to oral feeds and shorter hospital stay.

2.4.2. Gestation

The prevalence of intra-uterine foetal death in gastroschisis is 4.48 per 100. On risk stratification according to gestation age, the prevalence of foetal death for pregnancy that reaches beyond 36 weeks of gestation is 1.28 per 100 [8].

2.4.3. Surgical management

Surgical management is best described by dividing the gastroschisis into simple and complex groups.

2.4.3.1. Simple gastroschisis. Simple gastroschisis (Fig. 1a) is when the bowel is in good condition and is amenable to closure by means of primary surgical repair or application of a preformed spring loaded silo. The advantages of primary repair are immediate closure, decreased length of hospital stay, less intensive care bed days and less time to achieve full feeds [9]. In comparison, the preformed silo has been reported to decrease the risk of intraabdominal compartment syndrome, hence leading to a reduction in the requirement of prolonged ventilation, decrease in bowel ischaemia and reduced incidence of necrotising enterocolitis [9].

Complications from silo application are well described; the most frequent problem being dislodgement followed by difficulty in achieving fascial closure [10]. Intestinal injury can also occur and some surgeons feel that the long cylindrical column of the silo itself adds to a decreased bowel perfusion and the narrow neck of the silo base is a significant risk for volvulus and ischaemia if not managed appropriately [11]. These risks may be overcome by proper training to both medical and nursing

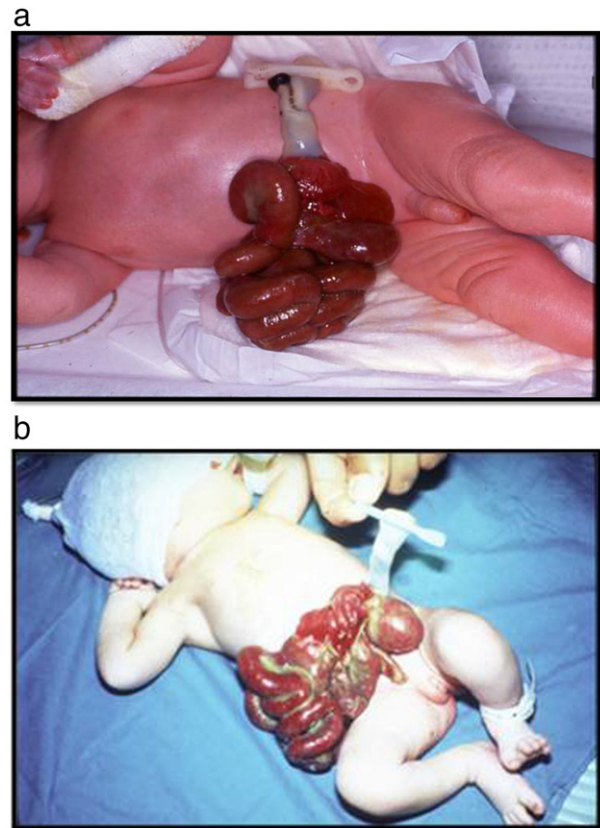


Fig. 1. a – Simple gastroschisis. b – Complex gastroschisis.

staff in the application of the silo and the follow on care [12]. The overall outcomes for both the surgical techniques are the same and use of preformed silo device should be in selective cases [13].

2.4.3.2. Complex gastroschisis. Complex gastroschisis (Fig. 1b) is when the bowel is either inflamed, adherent with peel, dilated, perforated, atretic, ischaemic or necrotic. Over 20% have complex gastroschisis which include intestinal atresia in two-third of cases and can be further complicated by gangrene, closing gastroschisis, perforation, strictures or volvulus. Primary closure may not be achieved in almost 50% of cases and these patients will require staged management with silo application to allow for bowel recovery and further staged bowel surgery in the form of a stoma formation, bowel resection and final bowel anastomosis for gastrointestinal continuity [14].

All patients with gastroschisis whether simple or complex will require central venous access for parenteral nutrition due to transient gut motility problems causing delay in enteral feeding.

2.4.3.3. Outcome. The overall survival rate up to one year is 96%. Mean hospital stay is around 53 days. Only 16% have a good course without any complications [15]. Type of primary reduction – either surgical or silo did not make any difference to the outcome [16]. The main problem during follow up is poor weight gain in about 30% of cases and remains at <10th centile at 1 year. Other complications include sepsis in 37%, necrotising enterocolitis in 10%, parenteral nutrition related cholestasis in 25% and short gut syndrome in 13% [17].

The mean duration of hospital stay with complex gastroschisis is double that of simple gastroschisis at 105 days. A third of this cohort of patients will go home on parenteral nutrition. Survival after complex gastroschisis is 89% and majority of patients achieve full oral feeding at 2 year followup [14].

The management of short gut remains a difficult aspect in the management of complex gastroschisis. Overall survival after short gut from

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