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REVIEW

Clinical characteristics and outcome of omphalocele and gastroschisis: a 20-year multicenter regional experience

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Abstract

Omphalocele and gastroschisis are the most common types of abdominal wall defects. Comprehensive local experience helps parents to make decisions on the pregnancy and foresee the disease journey. A retrospective review of abdominal wall defect patients in all three pediatric surgical centers in Hong Kong between January 2003 and February 2023 was conducted. All patients consecutively diagnosed with omphalocele and gastroschisis were included, excluding other forms. Data of demographics and short- and long-term outcome parameters were collected. A total of 99 cases were reviewed and 85 patients met the inclusion criteria. Diagnoses include omphalocele major ($n=49$, 57.6%), omphalocele minor ($n=22$, 25.9%) and gastroschisis ($n=14$, 16.5%), with mean gestational age 37 weeks (SD 2.2) and birth weight 2.7 kg (SD 0.6). Omphalocele is most commonly associated with cardiovascular ($n=28$, 39.4%) and chromosomal defects ($n=11$, 15.5%). Surgical procedures including primary repair ($n=38$, 53.5%), staged closure ($n=30$, 42.3%) with average 8.6 days (SD 4.7) of silo reduction, and conservative management ($n=3$, 4.2%) were performed. The mortality rate was 14.1% ($n=10$) and the complication rate was 36.6% ($n=26$). The majority of patients had normal intellectual development (92.5%) and growth (79.2%) on the latest follow-up. For gastroschisis, one patient (7.1%) had intestinal atresia. Surgical procedures included primary repair ($n=9$, 64.3%) and staged closure ($n=5$, 35.7%) with average 8 days (SD 3.5) of silo reduction. Complication rate was 21.4% ($n=3$), with one mortality (7.1%). All patients had normal intellectual development and growth. The mean follow-up time of this series is 76.9 months (SD 62.9). Most abdominal wall defects in our series were managed surgically with a good overall survival rate and long-term outcome. This information is essential during antenatal and postnatal counseling for parents.

Keywords Omphalocele · Gastroschisis · Abdominal wall defects · Outcome

Introduction

Omphalocele and gastroschisis are the most common types of abdominal wall defects with different clinical presentations, with prevalence at 2.6 and 4 per 10,000 births [1, 2]. Omphalocele is an abdominal wall defect located at midline with eviscerated organs covered by the sac, attributed to lateral fold development failure. It is commonly associated with chromosomal or other major system such as cardiovascular congenital defects. Different forms of omphalocele included cephalic fold defect as pentalogy of Cantrell, caudal fold defect as cloacal exstrophy and hernia into umbilical cord related to failed omphalomesenteric duct involution. Gastroschisis is related to the abnormal involution of right umbilical vein leading to weakening of body wall, always presented to the right side of the abdomen, commonly associated with intestinal atresia [3].

Abdominal wall defects might be diagnosed on antenatal ultrasound morphology scan. Literature reported pregnancy termination rate at 50% for omphalocele with a chromosomal defect and 30% for isolated omphalocele [4]. After such antenatal diagnosis, it is important for expectant parents to comprehend the disease outcome to make confident decisions on the pregnancy. For those only identified upon delivery, being able to expect the disease journey of these newborns is also of paramount significance to the new parents. The aim of this study is to establish the largest

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retrospective review investigating the outcomes of omphalocele and gastroschisis in Hong Kong.

Methods

A 20-year retrospective review on patients under 18 years old diagnosed with abdominal wall defects in all three tertiary pediatric surgical centers in Hong Kong between January 2003 and February 2023 was performed. All patients with the diagnosis of omphalocele and gastroschisis were included. Exclusion criteria were all other forms of abdominal wall defects such as umbilical cord hernia, cloacal exstrophy, etc. Data was retrieved using the clinical data analysis reporting system, a territory-wide patient database in Hong Kong.

Definition of omphalocele major in this study, i.e., "giant" omphalocele, included omphalocele defect size of 5 cm or more, or if more than half of the liver was herniated into the sac, as referenced from literature, although no standardized definition is available worldwide [5]. Defect not fulfilling this was referred to as omphalocele minor.

Demographics such as gender, gestational age, birth weight, delivery mode, perinatal conditions, as well as abdominal wall defect types, surgical procedures, complications, short-term and long-term outcomes, associated conditions, chromosomal defects, and physical and intellectual development were collected.

Statistical analysis was done with Microsoft Excel and SPSS (Statistical Package for the Social Science version 28.0.) with independent sample's *t* test and Fisher's exact test.

Results

A total of 99 patient records were reviewed. Eight-five patients were included (41 males, 44 females), with omphalocele ($n=71$, 83.5%) with major type ($n=49$, 57.6%) and minor type ($n=22$, 25.9%) and gastroschisis ($n=14$, 16.5%). (Table 1) Other types of abdominal wall defects identified including 11 cases of umbilical cord hernia and 3 cloacal exstrophy were not included in this current study. Over the years, the incidence rate is in a decreasing trend (Fig. 1), ranging from highest at 16.1 per 100,000 live births in 2004, to 3.0 per 100,000 live births in 2022, with data of annual live births collected from the Census and Statistics Department, The Government of the Hong Kong Special Administrative Region [6, 7].

Table 1 Demographics of patients with omphalocele and gastroschisis

	Total ($n=85$)	Types of abdominal wall defects					
		Omphalocele ($n=71$; 83.5%)	Omphalocele major ($n=49$; 57.6%)	Omphalocele minor ($n=22$; 25.9%)	*P* value (major vs minor)	Gastroschisis ($n=14$; 16.5%)	*P* value (omphalocele vs gastroschisis)
Gender							
Male	41 (48.2%)	34 (47.9%)	24 (49.0%)	10 (45.5%)	0.803	7 (50%)	1.000
Female	44 (51.8%)	37 (52.1%)	25 (51.0%)	12 (54.5%)		7 (50%)	
Mean gestational age							
	37.0 weeks (SD 2.18)	37.1 weeks (SD 2.21)	36.9 weeks (SD 2.27)	37.6 weeks (SD 2.27)	0.193	36.7 weeks (SD 1.70)	0.602
Mean birth weight							
	2.69 kg (SD 0.63)	2.73 kg (SD 0.65)	2.61 kg (SD 0.61)	3.03 kg (SD 0.67)	0.017	2.42 kg (SD 0.38)	0.114
Delivery mode							
Vaginal delivery	28 (32.9%)	21 (29.6%)	10 (20.4%)	11 (50.0%)	0.009	7 (50%)	0.219
Cesarean section	54 (63.5%)	47 (66.2%)	38 (77.6%)	9 (40.9%)		7 (50%)	
Elective	24 (44.4%)	19 (40.4%)	16 (42.1%)	3 (33.3%)	0.703	5 (71.4%)	0.416
Emergency	24 (44.4%)	22 (46.8%)	17 (44.7%)	5 (55.6%)		2 (28.6%)	
Apgar score							
At 1 min	7.25 (SD 2.14)	7.18 (SD 2.18)	6.93 (SD 2.19)	7.82 (2.07)	0.154	7.64 (SD 1.96)	0.516
At 5 min	8.76 (SD 1.61)	8.72 (SD 1.69)	8.50 (SD 1.87)	9.29 (0.92)	0.102	9.00 (SD 1.10)	0.599

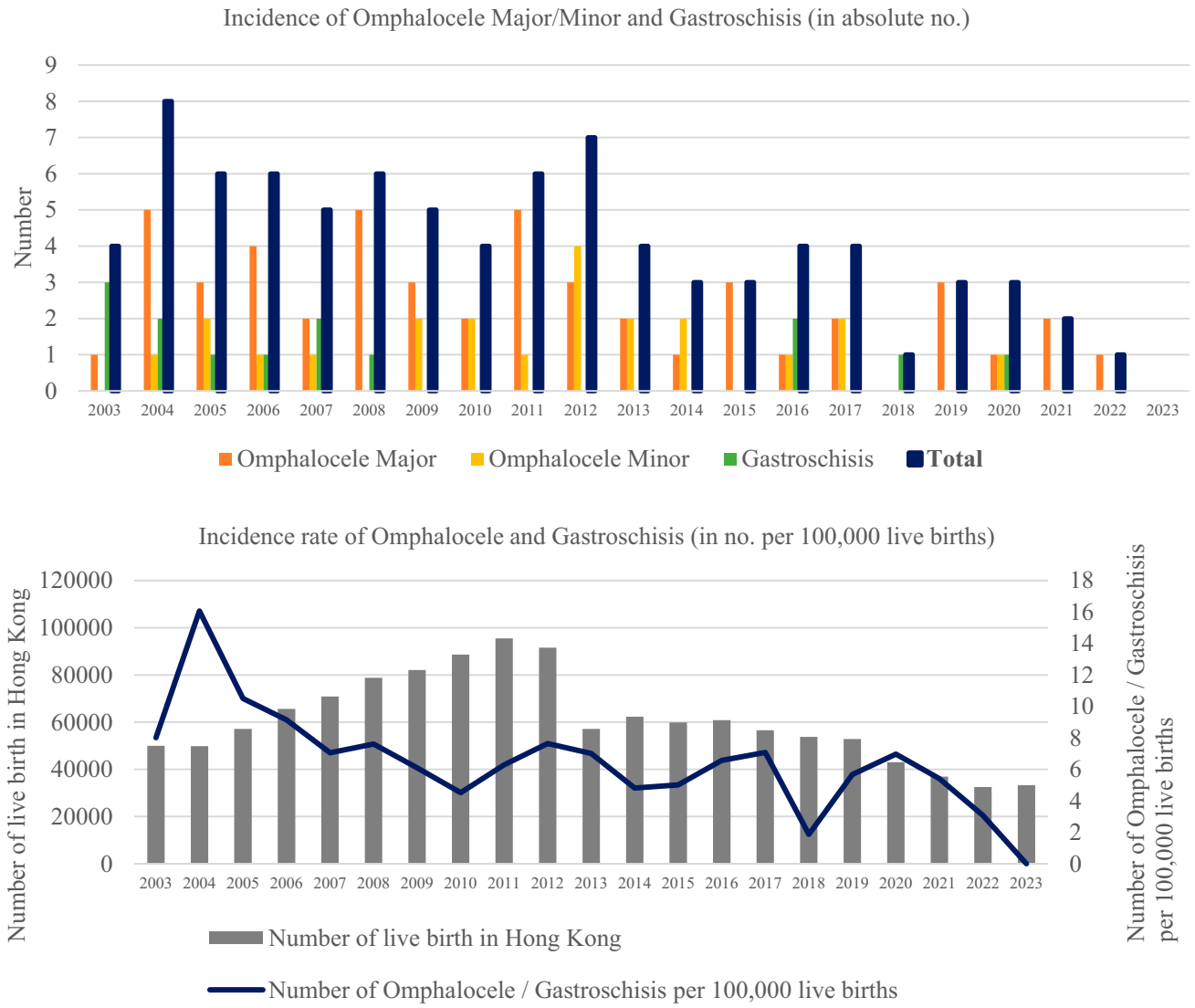

Fig. 1 Incidence of patients with ocentmphalocele and gastroschisis

Omphalocele group

Seventy-one patients were included, with 49 (57.6%) having major defects and 22 (25.9%) with minor. The mean gestational age was 37.1 weeks (SD 2.2) and birth weight was 2.7 kg (SD 0.7). Patients with major defect have a statistically lower birth weight (2.6 kg vs 3.0 kg, $p < 0.05$). Most patients with omphalocele major were born by cesarean sections ($n = 38$, 77.6% for major vs $n = 9$, 40.9%, $p < 0.05$). The mean Apgar score at 1 min was 7.2 (SD 2.2), and that at 5 min was 8.7 (SD 1.7). (Table 1).

For the surgical management of these patients, the strategies were in general the same among all three centers in Hong Kong. For primary closure, the sac was excised, with subsequent delivery of the eviscerated organs for inspection, and the abdominal wall muscle was undermined or stretched. Interrupted sutures were applied to the fascial layers with approximation of the two fascial edges with real-time observation of any increase in ventilation requirement. Fascial closure was followed by reconstruction of the umbilicus and closure of skin defect. Primary closure was achieved in 38 (53.5%) patients, up to 95.5% for the minor group. Thirty patients (42.3%) required staged closure. Decision of staged closure was commonly made after the abdominal cavity was deemed too small for full visceral reduction or when bedside reduction caused impairment of ventilation. After constructing the silo under general anesthesia, daily silo reduction was performed at bedside to reduce the protruded viscera until fit for fascial closure. The mean duration for silo reduction was 8.6 days (SD 4.7). One patient (1.4%) had biological mesh repair for fascia closure, whereas two (2.8%) had failed fascial closure and thus proceeded to skin

closure only. Three patients (4.2%) had non-surgical management, as two were not compatible with life with Patau syndrome, and one with omphalocele minor was not fit for general anesthesia and eventually had the abdominal wall defect spontaneously epithelialized (Table 2).

For short-term outcomes of the omphalocele group, early post-operative complications occurred in seven cases (9.6%), with post-operative bleeding ($n=5$, 7.0%), ileus with sepsis ($n=1$, 1.4%) and tension pneumothorax ($n=1$, 1.4%) (Table 3). The mean duration of weaning mechanical ventilation was 17.8 days (SD 48.9), with no statistical significance between the major and minor group (24.7 days vs 1.6 days ($p=0.09$). Mean time to starting enteral feeding was 11.7 days (SD 9.7) and was longer in the omphalocele major group (14.0 days vs 5.3 days, $p<0.05$). The duration of parenteral nutrition was longer in the major group (26.8 days vs 5 days, $p<0.05$). The mean length of stay was 141.3 days (SD 232.0) for major and 42.0 days (SD 41.3) for minor ($p=0.06$) (Table 4). Survival rate at 1 year was 85.9%. Ten patients (14.1%) did not survive, with details charted in Table 5. Among the group, five (7.0%) was associated with congenital heart conditions, 3 (4.2%) with Patau syndrome and two (2.8%) with severe sepsis. One (1.4%) of those patients with congenital heart conditions concomitantly had deranged liver function on parenteral nutrition with episode of upper gastrointestinal bleeding. One (1.4%) had massive small bowel necrosis. These ten mortality cases all occurred in the former half of this 20-year study period.

Long-term surgical complications occurred in 19 cases (26.8%) (Table 3). Of the two patients (2.8%) with initial skin closure, one was managed conservatively and another managed with delayed mesh closure. Eight other patients (11.3%) with initial fascial closure were complicated by ventral hernia, four of which were managed conservatively and another four had surgical repair at an average 22.3 months old (SD 14.1), with three using mesh for repair (Table 6). Seven patients (9.9%) had wound infections, with four (5.6%) involving the use of mesh, of which all developed wound infection requiring mesh removal and one (1.4%) developing enterocutaneous fistula managed as stoma eventually, with details charted in Table 6. Also, two patients (2.8%) had delayed adhesive intestinal obstruction and were both managed conservatively (Table 3).

Table 2 Management of omphalocele and gastroschisis patients

	Omphalocele ($n=71$)	Omphalocele major ($n=49$)	Omphalocele minor ($n=22$)	Gastroschisis ($n=14$)
Conservative	3 (4.2%)	2 (4.1%)	1 (4.5%)	0
Primary closure	38 (53.5%)	17 (34.7%)	21 (95.5%)	9 (64.3%)
Staged closure	30 (42.3%)	30 (61.2%)	0	5 (35.7%)
Duration of silo reduction	8.61 days (SD 4.73)		–	8.00 days (SD 3.54)

Table 3 Post-operative complications in omphalocele patients

	Proportion	Remarks
Early post-op	7 (9.6%)	
Bleeding	5 (7.0%)	
Required blood transfusion	1 (1.4%)	
Hemorrhagic shock requiring CPR	1 (1.4%)	Resuscitation performed, complicated with acute kidney injury and chronic kidney disease eventually
Ileus with sepsis	1 (1.4%)	Post-operative day 20 post-silo reduction and closure, presented with abdominal distension and sepsis, managed medically
Tension pneumothorax	1 (1.4%)	Resuscitation performed
Late	19 (26.8%)	
Delayed ventral hernia	10 (14.1%)	
Skin closure only initially	2 (2.8%)	One managed conservatively, one with surgical repair with mesh at age 5.5 years
Managed conservatively	4 (5.6%)	
Surgical repair	4 (5.6%)	At mean 22.3 months old (SD 14.1)
Wound infection	7 (9.9%)	
Involved mesh use	4 (5.6%)	All required mesh removal
Enterocutaneous fistula	1 (1.4%)	Managed as stoma
Delayed adhesive IO	2 (2.8%)	Both managed conservatively

Table 4 Outcomes of omphalocele and gastroschisis patients

	Omphalocele ($n=71$)	Omphalocele major ($n=49$)	Omphalocele minor ($n=22$)	p-value	Gastroschisis ($n=14$)
Short-term outcomes					
Duration of ventilation	17.8 days (SD 48.90)	24.7 days (SD 57.22)	1.6 days (SD 2.85)	0.094	8.5 days (SD 6.08)
Time to enteral feeding	11.7 days (SD 9.69)	14.0 days (SD 9.80)	5.3 days (SD 5.81)	0.001 (<0.05)	13.2 days (SD 8.33)
Length of Parenteral nutrition	20.0 days (SD 26.25)	26.8 days (SD 28.21)	5.0 days (SD 12.01)	0.008 (<0.05)	25.1 days (SD 13.10)
Length of stay	111.2 days (SD 199.66)	141.3 days (SD 231.97)	42.0 days (SD 41.34)	0.063	32.2 days (SD 17.50)
Follow-up duration	79.2 months (SD 64.01)	91.9 months (SD 62.79)	53.7 months (SD 60.14)	0.032 (<0.005)	66.2 months (SD 58.66)
Mortality in 1 year	10 (14.1%)	8 (16.3%)	2 (9.1%)	0.714	1 (7.1%)
Complications	26 (36.6%)	20 (40.8%)	6 (12.2%)	0.302	3 (21.4%)

Table 5 Mortality in omphalocele patients

Case no.	Age of death	Year of case	Gender	Type of abdominal wall defect	Cause of death
1	0 day	2007	F	Omphalocele major	Patau syndrome
2	5 days	2008	F	Omphalocele major	Patau syndrome
3	6 days	2008	M	Omphalocele major	Asso. with congenital heart conditions
4	7 days	2005	M	Omphalocele minor	Patau syndrome
5	11 days	2008	F	Omphalocele major	Asso. with congenital heart conditions
6	38 days	2005	M	Omphalocele major	Suspected line sepsis with respiratory distress syndrome and acute kidney injury
7	41 days	2004	M	Omphalocele major	Asso. with congenital heart conditions
8	50 days	2012	M	Omphalocele major	Asso. with congenital cardiac defects Initially had complete surgical reduction and achieved full enteral feeding, developed acute abdominal distension with exploratory laparotomy performed found marked intra-abdominal adhesion with midgut volvulus with massive SB necrosis from 2nd/3rd segment of duodenal up to 2 cm from IC valve and was decided for open and closed
9	5 months	2003	M	Omphalocele major	Repeated ankle sepsis, with chronic lung disease
10	10 months	2004	M	Omphalocele minor	Asso. with congenital heart conditions, also with deranged liver function on parental nutrition with episode of upper gastrointestinal bleeding

Omphalocele is commonly associated with other conditions, which could have implications on the long-term outcome of these patients (Table 7). These patients were followed up for an average 79.2 months (SD 64.0). Confirmed chromosomal defects or clinically suspected syndromes occurred in 11 neonates (15.5%), with diagnoses including Beckwith–Wiedemann syndrome ($n=5$, 7%), Patau syndrome ($n=3$, 4.2%), and 1 (1.4%) each with trisomy 18 mosaic, chromosome 15q duplication, and suspected Donnai–Barrow syndrome. Other than those with Patau syndrome, the others survived. Twenty-eight patients (39.4%) had cardiac abnormalities, with patent ductus arteriosus ($n=12$, 16.9%), patent foramen ovale ($n=6$, 8.5%), atrial septal defect ($n=8$, 11.3%), ventricular septal defect ($n=7$, 9.9%), valvular diseases ($n=3$, 4.2%), tetralogy of Fallot ($n=2$, 2.8%), malposition of heart ($n=2$, 2.8%), and aortic root dilatation ($n=1$, 1.4%), with details charted in Table 7. Among all, eight (11.3%) passed away. Four (5.6%) developed heart failure, with two able to wean off diuretics. Nineteen survivors (95%) with cardiac abnormalities ($n=20$) eventually had no remarkable heart failure manifestations. Fifteen patients (21.1%) developed long-term respiratory complications, with 9 (12.7%) developing chronic lung disease, 3 (4.2%) with diaphragmatic eventration or absence,

Table 6 Mesh use for closure in omphalocele patients

Case no	Year of case	Type of abdominal wall defect	Surgical management and complications and outcome
1	2004	Omphalocele major, with defect size 7 cm x 8 cm	Developed ventral hernia up to ~ 12 × 8 cm at 5 years-old with rectus defect up to pericardium, thus with dual-mesh hernioplasty performed, complicated with mesh dehiscence and wound infection 3 months later
2	2006	Omphalocele major, with defect size 10 cm	Residual ventral hernia after staged closure with silo; repair with Prolene mesh at 22 months old, complicated with exposed mesh and wound infection at 3, 9, and 11 years old. Wound eventually epithelialized
3	2015	Omphalocele major, eviscerating liver, spleen and small bowel	Inadequate abdominal wall closure with silo exchange till 2 months old, developed left diaphragmatic hernia, with thoracoscopic repair and diaphragm reinforced with biodesign dual mesh Abdominal wall defect eventually epithelialized after conservative management, later spontaneously developed enterocutaneous fistula since 4 months old, which progressed into prolapsed small bowel and managed as stoma since 1.5 years old
4	2021	Omphalocele major, eviscerating whole liver, spleen and most small bowel	Change of silo on D8, with fascia defect up to 4 × 10 cm, repair with Synovis Peri-Guard Pericardium Repair Patch on D22, with delayed wound infection with infected mesh and wound dehiscence since 8 months old, eventually had fascia closure and wound healing at 14 months old

2 (2.8%) with tracheobronchomalacia (one of who required full-day BiPAP), 1 (1.4%) with tracheostenosis with tracheoplasty done, and 1 (1.4%) with central sleep apnea on nocturnal BiPAP. Four (5.6%) passed away. Otherwise, 90.5% ($n = 10$) of the survivors with respiratory conditions ($n = 11$) were able to tolerate room air.

Among the survivors ($n = 61$), six patients (9.8%) had remarkable feeding problem in the long run. Gastrostomy was performed in four patients (6.6%) for poor oromotor performance, with fundoplication in two for reflux, and one able to wean off gastrostomy at age 4 years. For physical development, electronic records of growth curve were limited. Twenty-five patients (52.1%) were documented to have body weight initially below the 3rd percentile, 11 of which were associated with concurrent cardiac conditions, 9 with respiratory, and 3 with chromosomal defects. Fourteen patients (56%) showed catch-up growth. Thirty-eight patients (79.2%) achieved body weight above the 3rd percentile at the latest follow-up. As for intellectual development, seven (11.5%) had mental retardation or global development delay, with three having chromosomal defects. For those without chromosomal defects ($n = 53$), normal intellectual development was achieved in 92.5%.

Gastroschisis group

Fourteen patients were included with male and female accounting for 50% each. Mean gestational age was 36.7 weeks (SD 1.7) and mean birth weight was 2.4 kg (SD 0.4). Half were born by vaginal delivery, while another half by cesarean section. The mean Apgar score at 1 min was 7.6 (SD 2.0) and that at 5 min was 9.0 (SD 1.1) (Table 1). None had any confirmed chromosomal or significant congenital defects involving other major systems, one of which (7.1%) was associated with intestinal atresia with 60 cm small bowel and intact colon remaining after resection.

The surgical management of these patients was in general the same among all three centers in Hong Kong. For primary closure, the eviscerated bowels were cleansed with betadine and inspected and reduced into the abdominal cavity with extended fascia defect, followed by closure of fascia and skin with umbilicoplasty. It was achieved in nine (64.3%) patients, whereas the remaining five patients (35.7%) required stage closure. Similar to omphalocele, upon clinical judgment that complete reduction of eviscerated bowels was limited by small abdominal cavity or compartment effect to ventilation and hemodynamics, silo was sutured with daily bedside reduction, and eventually defect in the layers closed. The mean duration for silo reduction was 8.0 days (SD 3.5). (Table 2).

Post-operatively, one patient (7.1%) developed early adhesive intestinal obstruction and later developed abdominal distention and bowel perforation diagnosed radiologically, which was not resolved with bedside bilateral glove drain insertion and ran a downhill cardiopulmonary course. The patient eventually succumbed at 50 days old, being the single (7.1%) mortality case of this group. The mean duration of weaning mechanical ventilation was 8.5 days (SD 6.1). That of starting enteral feeding was 13.2 days (SD 8.3), with the length of parenteral nutrition being 25.1 days (SD 13.1). The length of stay was average 32.2 days (SD 17.5). (Table 4) This group of patients was followed up for

Table 7 Associated conditions and outcomes of omphalocele patients

	Proportion	Remarks
Confirmed chromosomal defects or clinically suspected syndromes	11 (15.5%)	Had surgeries for the abdominal wall defects performed and survived, except those with Patau syndrome
Beckwith–Wiedemann syndrome	5 (7.0%)	
Patau/trisomy 13	3 (4.2%)	Born with multiple major congenital deformities and passed away, respectively, on day 0, 5 and 7, respectively The former two were managed conservatively due to no surgical benefit The latter had bowel perforation from omphalocele minor, found to be perforated Meckel's diverticulum, with excision and omphaloplasty performed on day 0 of life
Trisomy 18 mosaic	1 (1.4%)	
Chromosome 15q duplication	1 (1.4%)	
Donnai–Barrow syndrome (suspected)	1 (1.4%)	
Cardiac	28 (39.4%)	
Patent ductus arteriosus	12 (16.9%)	2 passed away shortly after birth 1 had transient heart failure 1 required surgical closure 8 resolved spontaneously
Atrial septal defect	8 (11.3%)	4 resolved spontaneously 3 required surgical repair 1 associated with multiple cardiac abnormalities including aortic root dilatation with heart failure
Ventricular septal defect	7 (9.9%)	3 passed away shortly after birth 1 spontaneously resolved 3 required surgical repair
Patent foramen ovale	6 (8.5%)	5 resolved spontaneously 1 passed away shortly after birth
Valvular diseases	3 (4.2%)	2 resolved spontaneously 1 associated with multiple cardiac abnormalities including VSD and was repaired
Tetralogy of Fallot	2 (2.8%)	Both were involved in mortality cases
Malposition of heart	2 (2.8%)	Both were asymptomatic
Aortic root dilation	1 (1.4%)	Developed heart failure
Persistent heart failure	1/20 (5%)	3 had heart failure with 2 able to wean off diuretics on latest follow up
Respiratory	15 (21.1%)	
Chronic lung disease	10 (14.1%)	
Diaphragmatic eventration or absence	3 (4.2%)	
Tracheobronchomalacia	2 (2.8%)	1 required full-day BiPAP
Tracheostenosis	1 (1.4%)	With closure and tracheoplasty done
Central sleep apnea	1 (1.4%)	On nocturnal BiPAP
Tolerate room air	10/11 (90.5%)	
Long-term feeding problem	6/61 (9.8%)	
Gastrostomy	4/61 (6.6%)	2 with fundoplication 1 weaned off at 4 years old
Other feeding problem	2/61 (3.3%)	1 with chronic lung disease able to wean off nasogastric tube feeding at about 2 years old 1 with reflux and suspected functional partial intestinal obstruction able to wean off nasojejunal tube feeding at around 3.5 years old
Physical developmental problem		
Initial BW < 3rd percentile	25/48 (52.1%)	14 (56%) showed catch-up growth 11 associated with cardiac conditions, 9 with respiratory conditions, 3 with chromosomal defects
BW > 3rd percentile at latest FU	38/48 (79.2%)	

Table 7 (continued)

	Proportion	Remarks
Intellectual developmental problem		
Mental retardation or global developmental delay	7/61 (11.5%)	3 had chromosomal defects
Normal intellectual development in those w/o chromosomal defects	49/53 (92.5%)	

an average of 66.2 months (SD 58.7). The one patient (7.1%) with short gut after bowel resection with intestinal atresia was able to achieve full enteral feeding upon discharge at age 7 weeks, and two (14.3%) developed delayed adhesive intestinal obstruction at age 6 and 16 years, respectively, with the former managed conservatively, while the latter had surgical adhesiolysis and bowel resection and then fully recovered. All (100%) developed normally both physically and intellectually.

Discussion

Omphalocele and gastroschisis remain the most common forms of abdominal wall defects. Management options include primary repair, staged closure with initial silo placement and conservative management. Our study represents the first local multi-center long-term review of patients with abdominal wall defects.

Omphalocele is more likely to be associated with other comorbidities (39.4% with cardiac conditions and 15.5% with confirmed chromosomal defects or clinically suspected syndromes in our series), comparable with other literature reporting on 17% having chromosomal anomalies, with the most common ones being Beckwith–Wiedemann syndrome and trisomy 13, 18 or 21 [4], as well as 32% associated with congenital heart defects [8]. An overall survival rate of 75% was generally reported in the literature, and that if the associated anomalies were not present, the health status and long-term outcomes were even more favorable, similar to the gastroschisis group which was even less associated with chromosomal or other defects [9]. The survival rate in our series was compatible to published literature, with survival rate of the omphalocele group 85.9% and gastroschisis 92.9%. From our studies, all the ten mortality cases of omphalocele and the one mortality case of gastroschisis occurred in the former 10 years of this 20-year study period. All the three fatal Patau syndrome cases were born in 2005–2008. It is our postulation that the popularization and advancement in antenatal screening over the years aided in early diagnosis of associating congenital anomalies, especially fatal ones with increase in overall survival rates.

Earlier studies have concluded that normal physical and intellectual development can be expected, except with severe additional anomalies or chromosomal defects, and thus isolated omphalocele or gastroschisis is not an indication for termination of pregnancy [10]. There was a decreasing prevalence of abdominal wall defects in our locality over the study period, which we postulate to be due to a higher rate of antenatal diagnosis leading to a higher rate of termination of pregnancy. However, more data has to be retrieved to support our postulation.

In terms of management, most of our patients had operative management. Primary repair was achieved in 53.5% of patients with omphalocele, which was higher than 30%–50% quoted from existing studies [11]. The remaining 42.3% had closure after silo reduction. Only three cases (4.2%) had non-surgical management. This might be due to the fact that only one patient in our series was deemed unfit for general anesthesia and thus most of them could undergo surgical repair. In addition, Bauman et al. reported the side effects of different topical agents, including toxicity of mercurochrome, hyponatremia due to hypotonicity from silver nitrate, and thyroid hormone suppression by betadine [11]. Furthermore, complete neoepithelialization of the omphalocele sac was reported to take up to 2–3 months [12]. Therefore, surgical repair remains our first choice if the patient is deemed to be fit for general anesthesia during the neonatal period.

There was also a low rate of the use of synthetic mesh in our series. Skarsgard et al. reported complication rate of non-absorbable mesh, e.g., Teflon™, Prolene™ and Gore-Tex™ use to be as high as 25%, with a more favorable outcome and lower infection rate with biological mesh such as AlloDerm™, Strattice™, and Permacol™ [5]. Yet, our experience was limited as only four patients in our series had mesh use. However, all were complicated by post-operative infection requiring removal eventually.

Our study has several limitations. As we were only able to retrieve data of patients born with the defects, data was not available regarding the overall incidence of prenatal diagnosis and termination of pregnancy. Also, as the study was over a 20-year period in few pediatric surgical centers, there was no standardization of reporting the operative findings. There were also possible recall bias and missed-out data. A larger-scale regional prospective cohort studies, as well as including the quality-of-life survey, could have led to a more powerful review of the long-term outcomes of patients with abdominal wall defects. Also, further studies into the family

or maternal history of this group of patients might play a role in identifying significant or novel risk factors of the disease.

Conclusion

Most abdominal wall defects in our series were managed surgically with a good overall survival rate and long-term outcome. This information is essential during antenatal and postnatal counseling for parents.

Author contributions The authors confirm contribution to the paper as follows: Study conception and design: VC, JH Patient data recruitment: VC, AF, VW Data collection: VC Data analysis and interpretation of results: VC, JH Manuscript preparation: VC, JH Final review and confirmation for publication: All authors.

Data availability No datasets were generated or analysed during the current study.

Declarations

Conflicts of interest There were no conflicts of interests to declare. No specific grant was received for this research. Ethical approval was obtained from the institutional review boards of all institutes.

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