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REVIEW ARTICLE

Antenatally Diagnosed Surgical Conditions: Fetus As Our Patient

Kashish Khanna[1] · Anjan Kumar Dhua[1] · Veereshwar Bhatnagar[1]

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Abstract
In today's era of improved antenatal care and screening modalities (3D high-resolution fetal ultrasound, fetal magnetic resonance imaging, fetal echocardiography, maternal serum markers and fetal blood sampling), an early diagnosis of surgical fetal abnormalities is routinely possible. A thorough knowledge about the incidence of such defects, the means of detection and evaluation, the scope of fetal intervention, postnatal management and long-term outcomes of common surgical conditions diagnosed antenatally is essential. This knowledge would not only help in proper patient care and management but also to ensure appropriate counselling of the expectant parents. This article highlights the fetal anomalies which are amenable to some form of fetal intervention from a pediatric surgical perspective as also those which can be treated after birth.

Keywords Fetal surgical anomalies · Fetal intervention · Congenital · Malformations · Antenatal diagnosis

Introduction

About 3–6% of the neonates may be born with congenital anomalies despite the advancements in prenatal diagnosis, intervention and counselling [1, 2]. When a fetus is diagnosed with a congenital malformation in-utero, the common questions asked by the expecting parents are about the fetal outcome, whether the condition is surgically correctable, possible antenatal interventions, the timing of delivery, mode of delivery, role of postnatal intervention and the long-term sequelae of the given malformation. A multidisciplinary team consisting of the obstetrician, neonatologist, pediatric surgeon, radiologist, and the geneticist is required to deal with such situations. In between the two extreme options of medical termination of pregnancy (MTP) or continuation till term, lie the domain of new invasive therapies and an additional possibility of arranging for delivery at a tertiary care hospital where a multidisciplinary team is available for the immediate care of the newborn [3]. The authors have reviewed the common antenatally diagnosed surgical conditions (ADSC) encountered and its impact on fetal outcome.

✉ Veereshwar Bhatnagar
veereshwarb@hotmail.com

[1] Department of Pediatric Surgery, All India Institute of Medical Sciences, New Delhi 110029, India

Prenatal Diagnosis of Surgical Conditions

In 2010, Gupta et al. [1] reported that out of 8000 antenatal cases seen each year at authors' centre, 5–6% had congenital anomalies. Of these, 80% were terminated at the level of obstetrician and only 20% were referred to a pediatric surgeon for an opinion on surgically correctable condition [1, 4]. Ultimately, less than a quarter of these referred patients required surgery. In 8.8% cases, there was a discrepancy in the antenatal and the postnatal diagnosis [1].

A routine antenatal ultrasound (USG) performed at 18–20 wk of gestation can pick up most of the standard fetal anomalies. Mothers with high-risk factors like raised alpha-fetoprotein (AFP), family history of chromosomal abnormalities, advanced maternal age, previous spontaneous abortions or fetal demise, twin pregnancies *etc*. require an earlier scan between 10 and 13 wk. Some of the specific surgical conditions detected during prenatal screening have been discussed below focusing on those conditions which are amenable to antenatal interventions or are surgically correctable (Table 1).

Central Nervous System (CNS)

Common CNS anomalies include agenesis of corpus callosum, arachnoid cyst, cerebral calcifications, intracranial hemorrhage, macrocephaly, microcephaly and neural tube defects (NTD) like anencephaly, myelomeningocele (MMC) with or without hydrocephalus. Failure of closure of the neural

Table 1 Potential for fetal interventions in antenatally diagnosed surgical conditions

Fetal malformation	Fetal presentation	Fetal neonatal consequence	Treatable by
Shunts/Needle Aspiration			
1. Posterior Urethral Valves	Oligohydramnios Hydronephrosis	Renal dysplasia Renal insufficiency Pulmonary hypoplasia and respiratory insufficiency	Vesicoamniotic shunts
2. Congenital Cystic Adenomatoid Malformation (CCAM) of Lungs	Mediastinal shift Hydrops	Pulmonary hypoplasia	Thoracocentesis and thoracoamniotic shunting in cases of hydrops
3. Fetal Hydrothorax	Mediastinal shift Hydrops Polyhydramnios	Pulmonary hypoplasia	Thoracoamniotic shunt
4. Aqueductal Stenosis	Hydrocephalus	Neurologic damage	Ventriculoamniotic shunt
5. Ovarian Cysts	Abdominal mass Polyhydramnios	Ovarian torsion	Needle aspiration in case of large cysts, small cysts regress after delivery
Fetoscopic Surgery			
1. Twin-Twin Transfusion Syndrome (TTTS)	Monochorionic twin pregnancy, polyhydramnios with a maximum vertical pocket (MVP) ≥8.0 cm (with a distended bladder) in the recipient and oligohydramnios with an MVP ≤2.0 cm (with a non-distended bladder) in the donor	Hydrops IUFD	Fetoscopic laser photocoagulation
2. Congenital Diaphragmatic Hernia	Stomach seen in the thoracic cavity at the level of 4 chambered heart, the absence of stomach bubble in the abdomen, mediastinal shift	Pulmonary hypoplasia Pulmonary hypertension IUGR	Balloon occlusion of the trachea (FETENDO)
3. Posterior Urethral Valves	Oligohydramnios Hydronephrosis	Renal dysplasia Renal insufficiency Pulmonary hypoplasia and respiratory insufficiency	Cystoscopic ablation of valves
Open Fetal Surgery			
1. CCAM	Mediastinal shift Hydrops	Pulmonary hypoplasia	Lobectomy
2. Sacrococcygeal Teratoma	High output failure hydrops	IUFD Prematurity Hemorrhage	Resection
3. Meningomyelocele	Neural tube defect with or without hydrocephalus Arnold Chiari malformation	Spectrum from normal to severe neurological impairment of lower extremities, bladder and bowel	Repair
4. Cervical Teratoma	Neck mass	Life-threatening airway obstruction	Resection
Fetal Image-Guided Surgery (FIGS)			
1. Aortic Stenosis	IUGR Hydrops	Severe metabolic acidosis IUFD	Balloon dilatation
2. Anomalous Twins	IUGR	Preterm delivery	RFA of anomalous twins

IUFD Intrauterine fetal death; *IUGR* Intrauterine growth retardation; *FETENDO* Fetal endoscopic occlusion of trachea; *RFA* Radiofrequency ablation

tube during the first four weeks after conception can result in various forms of MMC. The incidence is about 1 in 1000 pregnancies [3]. A population-based survey of NTD births in rural areas of North India showed that the incidence was 7.48 per 1000 live births [5]. Mothers who are obese, on anticonvulsant medications, or have a short inter-pregnancy interval are at a higher risk. Preconception folic acid (at least 400 μg a day for three months) reduces the incidence of NTD.

Sonographic findings show a dorsal MMC sac, splaying of the dorsal vertebrae with or without associated lateral ventriculomegaly and Arnold Chiari-II malformation (ACM). In cases of ventriculomegaly (>11 mm width), a dangling choroid plexus can be observed on a transventricular view [6]. Furthermore, the frontal bones may lose their convex curvature and appear inwardly flattened (lemon sign). In a transcerebellar view, the banana sign due to the anterior curving of the cerebellum because of the small posterior fossa may be observed. These features are suggestive of ACM. An increased maternal serum AFP helps in early screening and confirmation of NTDs. A karyotyping to rule out associated chromosomal abnormalities is essential.

Fetal intervention (FI) started with the open fetal MMC repair first in animal models, followed by human fetuses. The management of meningomyelocele (MOM) trial was conducted to assess and compare the results of fetal MMC repair to neonatal surgery [7]. Total 183 eligible pregnant mothers were randomised and operated at three centres. A significant reduction in the need for shunt placement, reversal of hindbrain herniation, and improvement in functional motor level was observed in the prenatal surgery group with a trade-off of a significant number of premature deliveries due to chorionic membrane separation, spontaneous membrane rupture, preterm labour and oligohydramnios [7–9]. Though open fetal surgery is still being practised worldover post MOM's era, efforts to find newer minimally invasive approach (fetoscopic repair) are evolving [10].

Thoracic Anomalies

The most common thoracic abnormality is congenital diaphragmatic hernia (CDH) with an incidence of 1 in 3000 live births [2]. Others include bronchopulmonary sequestration (BPS), congenital pulmonary adenomatoid malformation (CPAM), hydrothorax, and pulmonary agenesis.

CDH is a defect in the diaphragm due to the failure of closure of the pleuropulmonary membrane by the 9th–10th wk of gestation. The more common type, Bochdalek hernia presents as a posterolateral defect on the left side more than the right. It is associated with a variable degree of pulmonary hypoplasia and pulmonary hypertension, which remains the most important prognostic factor for morbidity and mortality. CDH is diagnosed if the stomach bubble is not observed in its usual intra-abdominal location and the abdominal viscera is seen herniated into the thoracic cavity at the level of the cross-sectional view of the four-chambered heart on USG. Other findings include a mediastinal shift to the right and a decreased circumference of the abdomen. The modalities for assessment include high-resolution USG, fetal MRI, echocardiography and genetic testing between 20 and 24 wk [11]. Hedrick et al. reported that syndromic, familial, bilateral, associated with specific genetic abnormalities and CDH with severe congenital cardiac anomalies had poorer outcomes [11]. Specific prognostic markers have been studied to assess the severity of CDH and plan FI when necessary (Table 2) [11]. Open fetal surgical repair had a high fetal mortality. The concept of lung expansion by tracheal occlusion and the subsequent pushing of the abdominal viscera back into the abdominal cavity gave birth to the fetal endoscopic tracheal occlusion technique (FETENDO) [12]. Here the fetal trachea was occluded endoscopically using a clip/balloon/detachable balloon which was removed at the time of delivery or dissolved spontaneously. FETENDO provided no improvement in survival but resulted in more premature rupture of membranes (35–100%), preterm delivery (50–73%), and fetal demise (0–2%) [13, 14].

CPAM is characterised by the appearance of multiple cystic to solid (echo dense) lung lesions with their blood supply derived from the pulmonary circulation. In contrast, BPS has a systemic feeding vessel. Fetal MRI may sometimes be required to differentiate between the two. CPAM is more common in males and is usually unilobar [15]. Bilateral disease and hydrops fetalis (large cysts, often type III lesions, causing mediastinal shift and vena cava obstruction lead to hydrops) are poor prognostic factors and result in death in-utero or just after birth [16]. Delivery should be planned at term at a tertiary care centre, and cesarean delivery is not indicated for CPAM except for the standard obstetric indications.

In a few select cases, FI has been performed in the form of simple centesis of amniotic fluid, thoraco-amniotic shunt (in large macrocystic CPAMs that cause hydrops) and percutaneous laser ablation [2, 17]. Shunt surgeries are performed between 20 and 35 wk. In those patients found unsuitable for intervention (microcystic variety), a course of maternal steroids was found effective in arresting the growth of the CPAM [18]. In rare cases of large microcystic cases when steroids are ineffective, open fetal surgical resection has been performed as salvage therapy. In cases with hydrops having a CPAM volume ratio or CVR (calculated volume of the lesion to the fetal head circumference) >1.6–2.0, and significant respiratory distress at birth, an ex-utero intrapartum treatment (EXIT) procedure can prove life-saving [19].

Genitourinary Tract

Echogenic kidneys, multicystic dysplastic kidneys (MCDK), polycystic kidney disease, hydronephrosis, ureteropelvic junction obstruction (UPJO), posterior urethral valves (PUV), ureterocele, ectopic ureter, ambiguous genitalia, cloaca *etc*. are all bothersome congenital genitourinary anomalies.

Table 2 Various prenatal prognostic markers for assessment of severity in a fetus with congenital diaphragmatic hernia

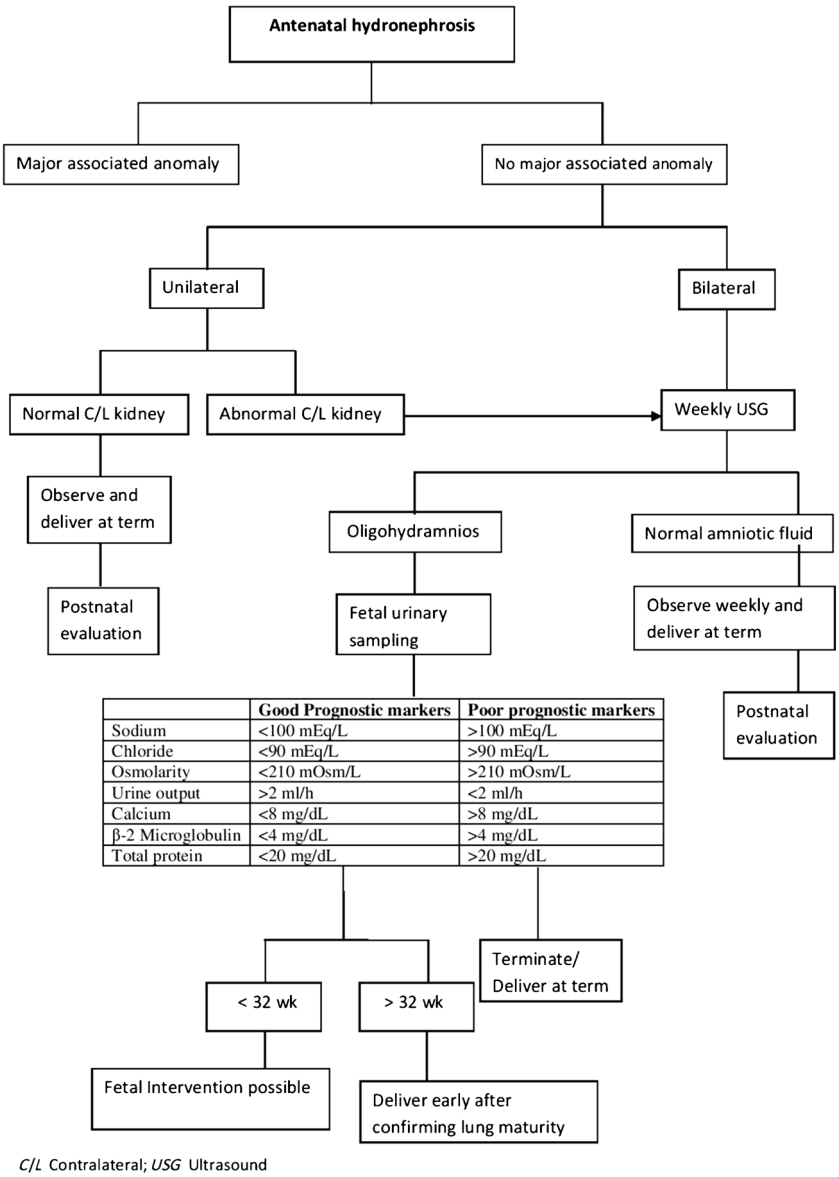
Prognostic Factor	Prognostic Groups	% Survival
Liver herniation	Liver up	65%
	Liver down	95%
Stomach herniation	**Grade 0**- Stomach intra-abdominal	93%-95%
	Grade 1- Stomach in left thorax	29%-41%
	Grade 2- Less than half of the stomach to right thorax	
	Grade 3- More than half of stomach to right thorax	
Liver herniation and stomach herniation	**Group 1-** Liver down	87%
	Group 2- Liver up, stomach grade 0-2	47%
	Group 3- Liver up, stomach grade 3	9.7%
Lung: Head Ratio (LHR) O/E LHR (%)	**Degree of pulmonary hypoplasia**	
	< 15- Extreme	Nil
	16-25- Severe	20%
	26-35, liver down- Moderate	30%-60%
	36-45, liver up- Moderate	
	36-45, liver down- Mild	>75%
	> 45 - Mild	
Fetal lung volume (FLV) O/E FLV (%)	<25	13%
	25-35	69%
	>35	83%

O/E Observed to Expected; % Percentage

Antenatal hydronephrosis (ANH) is diagnosed in 1% of pregnancies on USG [3, 20]. The common ANH grading systems are the Society for Fetal Urology (SFU) grading system and the anterior-posterior diameter (APD) of the dilated renal pelvis [21, 22]. Apart from the physiological fetal renal pelvic distension, a pathological ANH occurs when the APD is >10 mm [23]. If the APD is <10 mm, the prognosis remains excellent [2]. Spontaneous resolution is noted in 20% of the patients at birth and about 80% by three years [2]. Only 17% cases of ANH require surgery after birth [2] (Fig. 1).

PUV is the most common cause of bladder outlet obstruction (BOO) in males with an incidence of 1 in 2000 to 4000 live male births [2]. BOO predisposes the neonate to subsequent renal impairment. This may range from a completely asymptomatic pregnancy to complete obstruction leading to severe prenatal renal impairment with significantly associated oligohydramnios. When such oligohydramnios present between 16 and 24 wk, it is associated with pulmonary hypoplasia and resultant high perinatal fetal mortality and morbidity [24]. Long-standing high-grade obstruction may lead to Potter facies and clubfeet. The characteristic sonographic features of BOO (depending upon the severity) include marked and persistent dilatation of the urinary bladder, with thick and trabeculated bladder wall, dilated posterior urethra (keyhole sign), bilateral hydronephrosis and decreased amniotic fluid volume. High-grade BOO may present with fetal urinary ascites due to spontaneous decompression through rupture of the urinary tract [3]. Bilateral high-grade BOO may result in bilateral renal dysplasia seen as markedly increased renal parenchymal echogenicity (due to abundant fibrous tissue), and presence of subcortical cysts [25]. This differs from MCDK which shows the presence of multiple non-communicating cysts of variable sizes with normal renal parenchyma (interface) between these cysts. MCDK is often associated with contralateral urological anomalies and thus warrant postnatal evaluation. Infants with severe bilateral renal dysplasia, severe oligohydramnios and pulmonary hypoplasia are incompatible with life and do not warrant any intervention. Similarly, fetuses with bilateral hydronephrosis and normal amniotic fluid volume may not require any fetal intervention. None the less, repeated serial USG monitoring is required. Fetuses with good prognostic markers are candidates for FI by vesico-amniotic shunting (VAS) or by fetal cystoscopic ablation of urethral valve [25, 26].

The PLUTO study and randomised controlled trial were designed to study the effectiveness, cost-effectiveness and patient acceptability of VAS *vs*. the conservative management of



	Good Prognostic markers	Poor prognostic markers
Sodium	<100 mEq/L	>100 mEq/L
Chloride	<90 mEq/L	>90 mEq/L
Osmolarity	<210 mOsm/L	>210 mOsm/L
Urine output	>2 ml/h	<2 ml/h
Calcium	<8 mg/dL	>8 mg/dL
β-2 Microglobulin	<4 mg/dL	>4 mg/dL
Total protein	<20 mg/dL	>20 mg/dL

C/L Contralateral; *USG* Ultrasound

Fig. 1 Flowchart showing management strategy in a case of antenatally detected hydronephrosis

fetal lower urinary tract obstruction (LUTO) [24]. It was prematurely terminated after 31 women were randomised due to difficulty in recruitment. It was observed that VAS provided improved survival to 28 d and one year as compared to the conservative management, but it was not possible to prove benefit beyond reasonable doubt [24]. Although there was a notable improvement in the pulmonary function in the fetuses that underwent VAS, whether there is an improvement in renal function was unclear [25].

Congenital High Airway Obstruction Syndrome (CHAOS)

CHAOS is characterised by the presence of extremely large echogenic lungs, flattened diaphragms dilated tracheo-esophageal tree, ascites and features of non-immune hydrops (Fig. 2) [3]. This may be caused by laryngeal atresia, tracheal atresia or laryngeal cyst. Approximately one-third of the fetuses develop non-immune hydrops fetalis and die in-utero, one-third decompress *via* a tracheo-esophageal fistula, and the rest one-third may progress till 30–32 wk of gestation, and this one-third may call for an EXIT procedure [3]. Delivery should be planned at a tertiary care centre where the expertise and facilities for EXIT procedure are available. At authors' institute 4 EXIT procedures have been performed since 2008 for CHAOS lesions including one for a large cervical teratoma (Agarwala S, Personal communication/Unpublished data May 12, 2018).

Cystic hygroma is one such lesion where the key sonographic features include the presence of markedly enlarged nuchal translucency that extends along the entire fetal back with clearly visible septations. This should be differentiated from simple nuchal thickening (>3 mm) as cystic hygromas

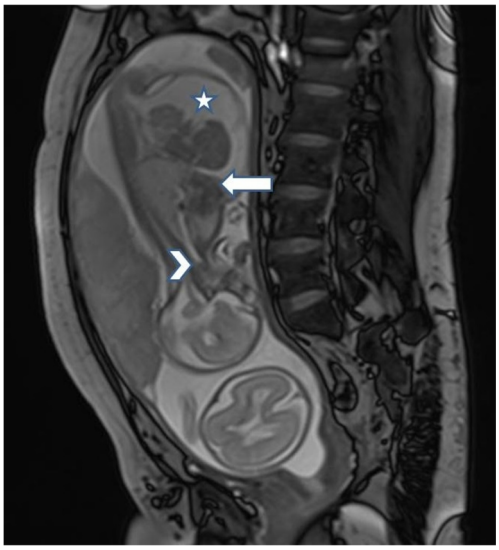


Fig. 2 Fetal MRI in twin pregnancy (32 wk gestation) showing one fetus with features of CHAOS. *Arrow* pointing at the inverted diaphragm, *arrow head* showing the abrupt cut-off of trachea till the subglottis and the star showing fetal ascitis. The postnatal diagnosis was congentital tracheal atresia

have a significant association with aneuploidy, cardiac malformations and fetal death as compared to the former [27]. Similar swellings along the anterior or anterolateral part of the neck presenting in the third trimester may be simple lymphangiomas which are not associated with other anomalies or hydrops. Cystic hygromas associated with non-immune hydrops are uniformly fatal [3].

Abdominal Wall Defects

Exomphalos is a ventral abdominal wall defect which is covered by a membrane (peritoneum and amnion) with an incidence of 1 in 4000 live births [2]. The typical sonographic feature of omphalocele is the umbilical cord insertion into the membrane covering a variable sized anterior defect in the fetal abdomen. In contrast, in gastroschisis, the umbilical cord inserts normally in the abdominal wall, and the defect usually lies to the right of the point of insertion. Also, in gastroschisis, the bowel appears to be freely floating, without a covering membrane with sometimes thickened or dilated bowel loops due to associated small bowel obstruction [3]. Exomphalos is associated with advanced maternal age, unlike gastroschisis which is commonly seen in infants of mothers <20 y [2]. Omphalocele can be a part of a syndrome like Beckwith-Weidmann (macroglossia, gigantism, exomphalos), Pentology of Cantrell (sternal, pericardial, intracardiac, diaphragmatic and abdominal wall defect), cloacal exstrophy *etc*. or can be associated with other cardiac, gastrointestinal and renal anomalies in 60–70% cases [2, 3]. Hence karyotyping is essential during prenatal evaluation. In a recent study by Lakasing et al., karyotyping was done in 380 cases of omphalocele; 250 had abnormalities, and 99% (248/250) of these fetuses died in-utero or underwent termination of pregnancy [28]. In case pregnancy is continued, serial sonographic monitoring for fetal growth restriction is necessary. Delivery by normal vaginal route at a tertiary care centre is advisable. In cases of large omphalocele (>5 cm), or cases with extracorporeal liver, a cesarean section is justified lest it should cause dystocia. Gastroschisis has a reported incidence of about 1.66–4.6/10,000 live births [2]. It has been associated with young maternal age, maternal smoking and amphetamine use. Fetuses have a high incidence of growth restriction, prematurity and third-trimester fetal demise; chromosomal abnormalities and extra-gastrointestinal abnormalities are not commonly associated. Vaginal delivery at a tertiary centre is recommended; cesarean section in cases of fetal distress is however justified [3]. Although efforts to test amnio-exchange to improve bowel function are going on, no current fetal intervention for gastroschisis is practical.

Gastrointestinal Abnormalities

Dilated loops of bowel (>15 mm in length and 7 mm in diameter) on prenatal USG are indicative of bowel obstruction. Pyloric stenosis (PS), duodenal atresia/stenosis, jejunal atresia, colonic atresia, Hirschsprung's disease and anorectal malformations are the common causes of intestinal obstruction. Antenatal USG shows a typical single bubble appearance in pyloric atresia/stenosis as opposed to double bubble in duodenal obstruction [3]. Dilated bowel, ascites, hyperperistalsis, polyhydramnios and echogenic bowel are soft signs on prenatal USG. In a meta-analysis, the detection rate of non-duodenal small bowel atresias by prenatal USG was highly variable, with values ranging from 10 to 100%, with an overall prediction of 50.6% (95% CI, 38.0–63.2%). When analyzed separately, the detection rates of jejunal and ileal atresia were 66.3%, (95% CI, 33.9–91.8%) and 25.9% (95% CI, 4.0–58.0%), respectively [29]. Polyhydramnios can be associated with more than half of these patients [3]. Serial USGs are needed in patients with duodenal atresia to rule out Down syndrome, which may be present in 30% of the cases. Amniocentesis for karyotyping and fetal echocardiography is recommended. There are no FI for intestinal atresias. For Hirschsprung's disease, diagnosis needs confirmation by a rectal biopsy after birth.

Miscellaneous Conditions

i) The common antenatally detected craniofacial anomalies include cleft lip and palate (CL/CP), macroglossia, micrognathia and agnathia, microphthalmia and athalmia. The overall incidence of CL/CP is 1 in 700 births [3]. Most cases are unilateral, commoner on the left side of the face and twice as common in males. These can be detected by transabdominal USG by 13–14 wk of gestation. Palatal defects can be better visualised by the 3D

Table 3 Postnatal interventions for antenatally diagnosed surgical conditions

	Condition	Timing of intervention[#]	Type of intervention
1	Congenital diaphragmatic hernia	Semi-urgent	Surgical repair of diaphragmatic defect
2	Congenital cystic adenomatoid malformation	Urgent/elective: depending upon the clinical symptoms	Lobectomy
3	Pyloric stenosis	Urgent	Pyloroplasty
4.	Duodenal atresia	Urgent	Duodenoduodenostomy
5.	Jejunal/ileal atresia	Urgent; Semi-urgent	Resection and anastomosis
5.	Tracheo-esophageal fistula	Urgent	Fistula ligation and repair of esophageal atresia
6.	Omphalocele	Urgent if causing intestinal obstruction; Elective when no associated obstruction	Repair or application of desiccating agent over sac, followed by excision of sac and repair of the defect later
7.	Gastroschisis	Urgent	Release of constricting ring and primary repair of defect; Silo creation if repair not possible followed by delayed closure of defect
8.	Anorectal malformations (ARM)	Urgent	High ARM- Colostomy; Low ARM males- Anoplasty, females- Anal dilatation
9	Neonatal Hirschsprung disease	Semi-urgent	Colostomy for decompression followed later by elective definitive repair or primary repair
10.	Posterior urethral valve	Urgent	Valve fulguration
11.	Uretero-pelvic junction obstruction	Elective	Pyeloplasty in symptomatic cases (pain/lump) or differential function <40% or progressive fall in function on repeated scans
12.	Exstrophy bladder	Elective	Single staged total reconstruction/ Modern staged repair of exstrophy
13.	Hypospadias	Elective	Hypospadias repair (single/ staged)
14.	Undescended testes	Elective (6–12 mo of age)	Orchidopexy
15	Choledochal cyst	Elective	Cyst excision and hepatico-duodenostomy/ roux-en –Y hepatico-jejunostomy
16	Extra-hepatic biliary atresia	Elective	Hepato-portoenterostomy
17	Sacrococcygeal teratoma (SCT)	Semi-urgent	Stabilisation followed by SCT excision with coccygectomy
18	Cervicofacial teratomas	Elective if airway is not compromised	Excision
19	Meningocele/ Meningomyelocele (MMC)	Semi-urgent/ Elective	Excision of sac and repair of MMC
20	Cleft lip	Elective (before 6 mo of age)	Cleft lip repair
21	Cleft palate	Elective (12–18 mo)	Cleft palate repair. Feeding advice to be followed till repair is done Orthodontic and speech therapy
22	Veno-lymphatic malformations	Elective (unless airway is compromised)	Surgical excision/ Sclerotherapy/ Pharmacotherapy
23	Congenital cysts (*e.g.*, branchial cysts/ thyroglossal cysts)	Elective	Excision

[#] Urgent = within 24–72 h; Semi-urgent = within the first 2 wk of life; Elective = later than 2 wk of life

USG [30]. MRI has a role to play in the evaluation of the secondary palate [31]. The presence of an abnormal karyotype and multiple associated anomalies can influence the decision to continue the pregnancy. Fetuses with an isolated CL/CP can normally be delivered at any hospital and can consult a pediatric surgeon after the birth of the infant. Due to the inherent properties of fetal skin and wound healing, scarless fetal lip and palate repairs may prevent postnatal scarring and the subsequent impaired facial growth [32, 33]. Till date, most researchers have focussed upon the restoration of CL/CP in animal models as fetal surgery for the non-life-threatening disease are still experimental till it becomes safe enough for the mother and fetus [34].

ii) Sacrococcygeal teratoma (SCT) is the commonest neonatal tumor, accounting for 1 in 35,000–40,000 births [2]. These tumors can be complex, solid and cystic masses in the sacrococcygeal region, but may have an intrapelvic or intra-abdominal extension. In large tumors, arteriovenous shunting may result in high-output cardiac failure. SCT >10 cm, substantial vascularity, more of solid components, rapidly growing tumor and a large tumor volume, have been associated with worse prognosis, hydrops and even fetal death [34–36]. Vaginal delivery is possible for small tumors. However, cesarean delivery is recommended to avoid trauma-induced hemorrhage or dystocia especially in large tumors >5-10 cm in size [3]. Early preterm delivery can be due to FI but also due to intrauterine fetal death, fetal cardiac failure, maternal mirror syndrome or polyhydramnios [37].

Fetal surgery for large SCTs when hydrops and fetal cardiac insufficiency are identified, especially at pre-viable gestational ages has been performed. Open fetal surgery with in-utero resection of the tumor is possible but, with high maternal and fetal morbidity due to a large incision on the uterus. Studies elucidating the minimally invasive approach to reduce the tumor blood supply and thereby reduce tumor size without causing tumor necrosis include laser ablation of the feeding vessel using low energy and power laser or ablation of the tumor itself [38].

Impact of Antenatal Diagnosis

Antenatal detection of surgical conditions certainly has an impact on the outcome. It provides an opportunity for appropriate

antenatal counselling, the conduct of timely MTP when indicated, screening for genetic syndromes, and ensures psychosocial preparedness on the part of the parents and the family. It helps in in-utero transfer of the fetus, ensuring an institutional delivery and planning the best mode of delivery (vaginal/cesarean). It also prepares the surgeon for planning an EXIT procedure in conditions like CHAOS. All ADSC should be confirmed by postnatal assessment and scans before planning any surgical intervention. Table 3 summarises the commonly performed postnatal interventions in a few of the common ADSC.

In an observational study conducted at authors' centre during 2014–2016, Sharma et al. [39] reported 68 patients who came with antenatal congenital surgical anomalies (CSA) for consultation as compared to 49 patients with CSA who reported during the postnatal period [39]. Out of 117 patients, 61.5% had urological or neurological anomalies (72), gastrointestinal (10), CDH (6), lung and chest anomalies (6), tumors (6), abdominal wall defects (4), intra-abdominal cysts (5), and others had rare CSA. Nine underwent MTP and 4 had an intrauterine fetal demise. Out of the remaining 55, two had no anomaly detected on the postnatal scan; 32 were managed conservatively; 1 denied surgery (conjoint twin) and the rest 20 were advised surgical management. In the latter group, three died before surgery, five after surgery and the rest 12 survived post-surgery. The authors concluded that appropriate prenatal counselling ensured proper perinatal care [39].

In a retrospective study by Raboei et al. [40], 234/400 fetuses were diagnosed antenatally with surgical anomalies namely: genital-renal (146; 58.2%), gastrointestinal (13; 5.2%), head and neck (7; 2.8%), gastro-pelvic (7; 2.8%), cardio-gastric (5; 1.9%), neuro-gastric (3; 1.2%), neuro-renal (11; 4.4%), cardio-renal (16; 6.4%), gastro-renal (7; 2.8%), cardiothoracic (9; 3.6%) and fatal (15; 5.9%). Associated anomalies were found in 92 cases [40]. The pediatric surgeon was the main team member who took the decision in groups involving amniocentesis for karyotyping (83/400) and those involving termination of pregnancy (26/83). The perinatal management was altered in 75% of cases. Nef et al. [41] conducted a study to assess the outcome after prenatal diagnosis of congenital anomalies of the kidney and urinary tract (CAKUT). They divided their cohort of 115 patients into low risk (unilateral nephrouropathy/bilateral isolated pelvic dilatation/normal amniotic fluid) and high-risk groups (bilateral pelvic dilatation and renal anomalies/ suspected PUV/ oligohydramnios/extrarenal anomalies). During the postnatal long-term follow up (median 2.2 y, range 0.1–18 y), they found that one-third each showed normalization, need of surgery or persistence of anomalies without the need of surgery [41].

A recent French study had compared prenatal and neonatal characteristics and outcomes in children with prenatal diagnosis (75) of esophageal atresia (EA) type A to those with a postnatal diagnosis (13) until the age of one [42]. They concluded that even though prenatal diagnosis leads to antenatal parental counselling, better antenatal management, and avoided postnatal transfers, however, it did not change the mortality rate or the one-year morbidity rate in infants diagnosed prenatally with pure EA.

Conclusions

Prenatal care of mother carrying a fetus with surgical malformation involves the care of the mother and the care of the fetus as our patient. It becomes the duty of the obstetrician, pediatrician, pediatric surgeon, radiologist and the family to collectively understand, support and treat both the mother and the fetus. Some conditions like CDH, MMC and LUTO can be dealt with in-utero, and the others like TEF require immediate postnatal intervention. However, certain conditions like bilateral renal agenesis, multiple congenital anomalies with abdominal wall defects and ruptured MMC or rachischisis may require termination of pregnancy. Antenatal diagnosis ensures better perinatal care and in-utero transfer of infants with ADSC.

Authors' Contributions KK: Collection of data and preparation of draft manuscript; AKD: Preparation of manuscript; VB: Conceptualising, planning, finalising the paper and will act as guarantor for this paper.

Compliance with Ethical Standards

Conflict of Interest None.

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