Case Report

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Situs inversus in association with duodenal atresia

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ABSTRACT

Situs inversus in association with duodenal atresia is very rare. A high index of suspicion coupled with appropriate evaluation is necessary for diagnosis and operative planning. We report a case of a 5-day-old who presented with duodenal atresia associated with polysplenia and situs inversus with a review of the medical literature.

Key words: Situs inversus, duodenal atresia

INTRODUCTION

Situs inversus is rare and confusing and it causes mirror image positioning of abdominal and thoracic viscera with incidence of about 1 in 10,000 of the normal population.[1] Gastro-intestinal and cardiac anomalies associated with situs inversus are very rare.[2]

Duodenal atresia is estimated to appear with incidence of about l in 4,000 to l in 15,000 live births and almost half of them have an associated congenital anomaly of other organ system.[1]

Association of duodenal atresia with situs inversus is rarely reported in the literature. There are about 20 patients reported in the literature having situs inversus in association with duodenal atresia.[2]

We report another case of duodenal atresia in association with situs inversus to highlight the importance of the reverse double bubble sign as a means of identifying this condition and the significance of keeping the mirror image positioning in mind when carrying out surgical operation.

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CASE REPORT

A 5-day-old female neonate delivered at 32 weeks and 3 days of gestation through an emergency lower segment caesarean section on account of foetal distress by an un-booked 23 year old Para 3+0. She was earlier being managed by the neonatologist for severe birth asphyxia before referral.

Prenatal obstetric scan had revealed poly-hydramnios and weak foetal cardiac activity, and the antenatal period was uneventful.

Mother is not a known diabetic. The baby presented with 3-day history of bilious emesis and progressive abdominal distension. Baby passed meconium after birth. On examination, the neonate weighs 1.65 kg, pink at room air, anicteric and was well hydrated. The respiratory rate was 66 cycles//min with vesicular breath sounds. The heart rate was 116 beats /min. First and second heart sounds were heard without cardiac murmurs. The abdomen was distended, and the anus was patent. Baby was commenced on intravenous fluid and antibiotics with a urethral catheter passed, and a nasogastric tube inserted drained copious bilious effluent. The haematologic as well as blood chemistry were with normal range. The baby gram, Figure 1 revealed "reverse double bubble" sign with levocardia, which was thought to be due to wrong labelling on the side of the film. Echo-cardiograph was not carried out. The neonate had laparotomy. Intra-operative findings were: Stomach and polysplenia on the right side while the liver and gall bladder were on the left.

The caecum and appendix were located in the left lumbar region. Type I post-ampullary complete duodenal atresia with Ladd's band was also found.

Duodeno-duodenostomy with appendicectomy and release of Ladd's band were performed.

Patient did well until the sixth post-operative day when she suddenly suffered apnoeic attacks and died after failed attempt at resuscitation.

DISCUSSION

The incidence of duodenal atresia ranges from 1 in 4,000 to 1 in 15,000 live births with almost half of them having associated congenital anomalies of other organ systems. Down syndrome is the most common associated anomaly. The other common association are cardiac anomalies, annular pancreas and mal-rotation of the gut.[1,3]

The aetiology of duodenal atresia is unknown; however, embryonic failure of recanalization after the stage of solidification by rapidly growing epithelium between the 8th and 10th week of embryonic development was thought to result in the development of duodenal atresia.[4]

Situs inversus is rare with prevalence of l in 10,000 of the general population.[1] Genetics and embryology of situs inversus are complex and multifactorial. Familial inheritance has been reported in some patients.[1,2] Primary ciliary dyskinesia, an autosomal recessive disorder have been linked with situs anomalies in about 3-5% of patients. It is postulated that most heterotaxic disorders are multifactorial in their causation.[5]

The common conditions associated with situs inversus are serious congenital cardiac defects and splenic anomalies. Congenital cardiac anomalies are present in about 5-10% of patients with situs inversus.[2]

In the presented case, there was no facility (that is, echocardiogram) to determine if there was an underlying cardiac anomaly, which may have contributed to the demise of the patient. Our clinical examination; however, did not reveal any murmurs. Other common conditions associated with duodenal atresia include annular pancreas, pre-duodenal portal vein, duodenal stenosis or diaphragm or atresia.[1,3] The only other associated anomaly found in our patient is polysplenia.

The association of duodenal atresia with situs inversus is extremely rare.[1-3] A review of the English medical literature revealed that only 25 cases of this association had previously been reported including our own case confirming its rarity, [Table 1].

There was no sex predilection. The male to female ratio was almost equal. In most cases, the liver was on the left side with mirror image positioning of the small and large intestine. The spleen was found situated on the right in most of patients as in our patient.[6-11] There were 5 cases of polysplenia. In the index case presented, we also found polysplenia on the right, Figure 2. The stomach was on the right with mirror image positioning of the colon.

The various causes of duodenal obstruction include pre-duodenal portal vein, duodenal diaphragm, stenosis, and atresia. Most of the patients had complete atresia, requiring duodeno-duodenostomy.

Our index patient had post-ampullary complete duodenal atresia in the second part of the duodenum with Ladd's band on the left. The stomach was also on the right Figure 3.

Situs inversus can be asymptomatic and found incidentally during laparotomy for a different pathology



Figure 1: The reverse double bubble sign on the plain abdominal radiograph of the baby

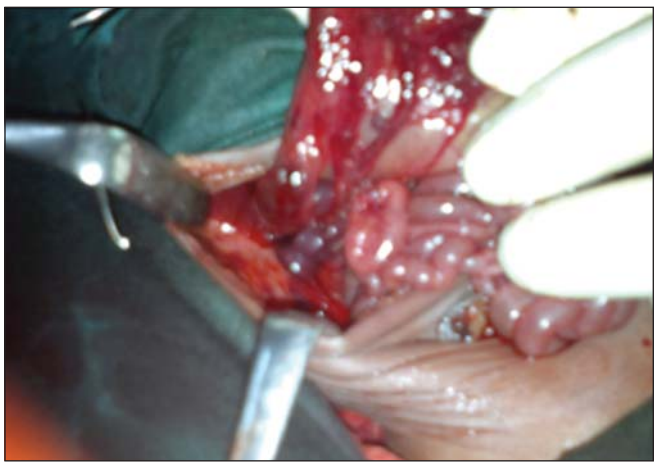


Figure 2: Associated polysplenia in the patient

Table 1: Duodenal atresia and situs inversus patients reported in the literature

Study	Sex	Presented	Spleen	Duodenum and colon	Heart	Surgery performed
1. Lee *et al.*	*	*	*	Duodenal atresia	*	Duodeno-jejunostomy
2. Nawaz *et al.*	F	2 day	Right	Malrotation of bowel	Fallot's tetralogy features	Side – to-side duodeno-duodenostomy+appendicectomy
3. Nawaz *et al*.	F	4 day	Right	L side, duodenal diaphragm with central hole	Normal	Duodenostomy, side – to – side duodeno – duodenostomy, appendicectomy
4 Iuchtman *et al*.[6]				Duodenal diaphragm Annular pancreas Internal hernia	Levocardia	"
5.Brown *et al*.[2]	F	Birth	Right (polysplenia)	Duodenal atresia	Interrupted IVC	Duodeno-duodenostomy
6.Park *et al*.[7]	F	6 day	Right	Doudenal atresia	Dextrocardia	Side to side duodeno-duodenostomy
7. Akel *et al*.	F	6 day	"	Duodenal obstruction – mucosal diaphragm	"	Duodenoplasty+appendicectomy
8.Sharma *et al*.[1]	M	11 day	Right	Duodenal diaphragm and Ladd's band	Dextrocardia, ASD	Duodestomy
9. Ziv *et al*.[8]	"	7 day	"	Duodenal atresia preduodenal portal vein colon on the right	Dextrocardia	Side to side duodeno-duodenostomy
10.Yasunori *et al*.[9]	M	1 day	"	Duodenal atresia, right sided mirror image colon	Levocardia	Duodeno-duodenostomy
11. Shankar *et a*l.[3]	M	2 day	Right	Caecum and appendix on the left Type I duodenal atresia, apple peel jejunal atresia	Levocardia, hypoplastic left ventricular double outlet ventricle	Side-to-side duodeno-duodeno-stomy
12. Lopez *et al*.[10]	-"	-"	"	Duodenal stenosis	Dextrocardia	-"
13. Lopez *et al*.[10]	"	"	Asplenia	Duodenal atresia ivermark syndrome	-"	"
14. Habib *et al*.[11]	"	"	Right	Duodenal atresia	Dextrocardia	Laparoscopic duodeno-duodenostomy
15.Chandran *et al*.	F	3 day	Right	Complete obstruction 2nd part. Duodenal atresia	Levoverted, IVC L	Duodenoplasty
16. Cacclaguerra *et al*.	M	1 year	Right polysplenia	Left sided, Ladd's bands across 2nd part. Mirror image of 180° malrotation	Normal	Duodenoplasty
17. Ohno *et al*.	M	27 weeks Gestation	Right polysplenia	Duodenal stenosis, colon iying on 2nd part of duodenum	Dextrocardia+cardiac abnormalities	Duodeno-duodenostomy, Blalock – Taussig shunt, Glenn and Fontan procedures
18. Adeyemi et al	F	7 day	Right	Duodenal atresia, ring of pancreatic tissue surrounding 2nd part	*	Side-to-side duodeno-jejunostomy
19. Adeyemi etal	M	12 day	*	Duodenal atresia, annular pancreas around 2nd part of duodenum, mucosal diaphragm	Normal	Duodenoplasty diaphragm excision
20. Adeyemi et al	M	8 day	Right	Duodenal atresia, complete annular pancreas surrounding duodenum	Normal	Side-to-side duo-deno-jejunostomy
21. Ruben *et al*.	*	*	Right polysplenia	*	Cardiomegaly, AV canal, absent IVC, L aortic arch	Mirror image porto-enterostomy
22.Fonkalsrud *et al*.	*	*	*	4 patients with SI, 2 duodenal atresia, 2 duodenal web	*	Unknown
23. Noack *et al*.	F	Birth	Asplenia	Situs ambiguous, lvermark syndrome, duodenal stenosis	Complete AVSD	Unknown
24. Tryfonas *et al*.	M	1 day	Right	Duodenum obstructed, R-sided colon mirror image non-rotation	Levocardia	Side-to-side duo-deno-jejunostomy+appendicectomy
25.Talabi *et al*. present series	F	5 day	Right polysplenia	Duodenal atresia, left sided Ladd's band across 2nd part, mirror image colon	Levocardia	End to end duodeno-duodeno-stomy Appendicectomy, release of Lladd's band

*Information not available. L=Left, R=Right, IVC=Inferior vena cava, AV=Atrioventricular, AVSD=Atrioventricular septal defect, SI=Situs Inversus; ASD=Atrial Septal Defect Numbers: 1, 2. 3, 7, 15, 16, 17, 18, 19, 20,21, 22, 23, 24 were cited from a table by Brown et al[2]



Figure 3: The stomach located on the right side with atresia of the 2nd part of duodenum

or at autopsy, but when associated with duodenal atresia it often presents early in the new-born period.[1] On the other hand, patient may present late.[12]

The diagnosis of duodenal atresia with situs inversus can be made easily with the high index of suspicion on plain babygram when a "reverse double bubble" sign is seen with a reverse heart shadow [Figure 1]. In our patient, the reverse double bubble sign was confusing. We thought initially that it was a case of wrong labelling of the side on the radiograph as it was the more common occurrence. Indeed, the film was wrongly labelled until it was reviewed with the radiologist and corrected. Though we eventually made the diagnosis of duodenal atresia, the associated situs inversus was missed and confirmed only at laparotomy.

The heart is the most common affected intra-thoracic organ *in situs* anomaly, and in most instances cardiac symptoms are usually the first ones that lead to the detection of this anomaly. It is important to establish the anatomy of the heart with an echocardiogram as part of the pre-operative assessment of the patient before undergoing any operative procedure so as to detect the presence of an associated cardiac anomaly, which may influence the prognosis of the patient.[2] In the index case presented, echocardiograph was not carried out as we missed the diagnosis of situs inversus pre-operatively.

The treatment of duodenal atresia with or without situs inversus is the same. The common ones are duodenotomy, duodeno-duodenostomy, and duodeno-jejunostomy. If the obstruction is due to duodenal diaphragm, excision with duodenoplasty is carried out. However, duodeno-duodenostomy is preferred by some surgeons so as to avoid bile duct injury due to the closeness of the ampulla of Vater to the duodenal diaphragm.[1]

The cause of death in our patient is unclear as all appeared to be going on well until the 6th day post-operative. Reviews of the literature suggest that the outcome depends on the severity of the associated cardiac defect. Echocardiographic detection of cardiac defect coupled with appropriate management by cardiac surgeon where available is associated with better prognosis. Lack of echocardiographic detection of the presence or absence of cardiac defects as in our index patient or where there is no expertise to manage associated cardiac anomaly, the outcome is poor.[2,3] Although, we made an attempt to get an autopsy carried out to unravel the actual cause of death, the parents declined.

In conclusion, duodenal atresia in association with situs inversus is very rare, a high index of suspicion is necessary especially when a reverse double bubble sign is seen on babygram. An echocardiograph should be carried out to define the anatomy of the heart before embarking on any surgical procedure. The outcome depends on the severity of associated cardiac anomaly.

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