

# Congenital Lumbar Hernia Associated with Duodenal Atresia and Lumbo-costo-vertebral Syndrome

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## ABSTRACT

Congenital lumbar hernia is associated with a multitude of congenital anomalies involving various other organ systems of the body. These involve the ribs, spine, muscles, kidneys, and spinal meninges. We are reporting

a case of one day old male child with congenital lumbar hernia associated with duodenal atresia and lumbo-costo-vertebral syndrome. We failed to find this association in literature. This prompted us to report this rare case.

**Keywords:** Hemivertebrae, Neonate, Scoliosis

## CASE REPORT

A one day old male child, born at 36 weeks gestation, was admitted to our Pediatric Surgery ward with complaint of a large mass occupying right lumbar region with non bilious vomiting. Baby belonged to a family with rural background. Mother was a house wife. She was non smoker and non-alcoholic, with no history of any harmful drug or radiation exposure during pregnancy. She was primigravida and never went for any antenatal check-up but the antenatal period was uneventful. Baby was delivered via normal vaginal delivery with a birth weight of 2 kg. Baby cried immediately at birth and no resuscitation was required. On physical examination, baby's vitals were stable. He had a length of 48 cm, head circumference of 32 cm. A mass of 5x6 cm was present in right lumbar region which was very soft and reducible by pressure, but increased enormously in volume when the child cried [Table/Fig-1,2]. After an informed consent from the parents, the baby was investigated. Infantogram of the baby revealed absence of lower ribs on right side with scoliosis of spine with hemivertebrae in thoracic region and duodenal atresia with massively dilated stomach and first part of duodenum showing double bubble sign



**[Table/Fig-1,2]:** Right lumbar hernia in a neonate.

**[Table/Fig-3]:** Infantogram showing absence of lower ribs on right side with scoliosis of spine with hemivertebrae in thoracic region and double bubble sign.

[Table/Fig-3]. USG was suggestive of lumbar hernia with normal liver, kidney and spleen but its picture could not be taken. Haemoglobin was 14 gm/dl and renal function tests were within normal limits.

Patient was put on nasogastric aspiration, i/v antibiotics and i/v fluid. After pre-operative resuscitation, patient was taken up for exploratory laparotomy. Operative findings were massive dilatation of stomach and first part of duodenum with type 1 duodenal atresia. Duodeno-duodenoplasty was done. After surgery, the patient was kept on i/v fluid and i/v antibiotics for 5 days. Once the GI aspirates decreased and patient started passing stool, he was started on feeds and was discharged on 10<sup>th</sup> day in stable condition. On follow-up child was well. Repair of hernia will be done after 6 months.

## DISCUSSION

Congenital hernias in the lumbar region account for only 10% of all the lumbar hernias. Less than 50 cases of congenital lumbar hernia associated with other congenital anomalies have been reported in English literature making it a rare entity [1, 2].

Congenital lumbar hernias usually appear as large compressible soft-tissue mass in the lumbar region. The 12<sup>th</sup> rib above, the iliac crest below, the vertebral spine medially, and the extreme oblique muscle laterally form the boundaries of lumbar region. The lumbar hernial sac may be empty, or may contain spleen, omentum, small bowel, appendix and colon, retroperitoneal fat, the bowel, kidney, omentum, colon, stomach, ovary, spleen, appendix and kidney. Lumbar hernias may be congenital or acquired (spontaneous, post traumatic, post operative, a post-regional suppurative process) [1,3]. The congenital form may be isolated, although it often occurs in association with the lumbo-costo-vertebral

syndrome, which includes one or more of following anomalies: Hemivertebra, rib abnormalities (absence, hypoplasia, fusion, gap), aplasia of dorsolumbar muscles, scoliosis. Other associated abnormalities reported include absence of the right kidney, eventration of the right hemi diaphragm, inguinal hernia, malrotation of the gut, ARM etc. But association with duodenal atresia has never been reported previously. X- ray, CT and US of the lumbar region may detect air filled bowel loops and solid organs in the sac [4].

Duodenal atresia is the congenital absence or complete closure of a portion of the lumen of the duodenum. The 3 anatomic types of duodenal atresia as described by Gray and Skandalakis are:

- **Type 1:** The most common type is formed by a membrane composed of mucosa and submucosa. This membrane traverses the internal diameter of the duodenum. The duodenum and stomach proximal to the obstruction are dilated and hypertrophied. The duodenum distal to the obstruction is narrowed.
- **Type 2:** The atretic ends of the duodenum are connected by a fibrous cord.
- **Type 3:** Complete separation of the atretic segments occurs [5].

The diagnosis of duodenal atresia is also usually confirmed by radiography. The treatment of lumbar hernia is surgical and should be performed in the first year of life [6,7]. Prognosis is good and recurrence is rare [7]. Early treatment of duodenal atresia includes removing fluids from the stomach via a nasogastric tube, and providing fluids intravenously [8]. Surgery (duodenoduodenostomy) is the definitive treatment, which may be performed openly or laparoscopically [9]. Prognosis is usually very good [8]. The initial repair has a 5 percent morbidity and

mortality rate [10]. Late complications may occur in about 12 percent of patients, and the mortality rate for these complications is 6 percent [10].

## CONCLUSION

Congenital lumbar hernia associated with other congenital anomalies is a rare entity. In this case we found duodenal atresia and lumbo-costo-vertebral syndrome as the associated anomalies. This association has never been reported in literature so far, making it probably the first such case to be reported.

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