

LUMBOCOSTOVERTEBRAL SYNDROME (LCVS) - A RARE ENTITY

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CASE SUMMARY-

Full term female neonate, weighing 2.7 kg, was born to 37-year-old mother with an uneventful antenatal course. On examination, child was active and anthropometric parameters were normal. Head to toe examination revealed a globular swelling below left costal margin lateral to dorsolumbar spine measuring approximately 6 cm X 6 cm. It was soft, nontender, reducible and used to become prominent on crying. Rest of general and systemic examination was normal. X-ray of chest and spine showed dysplastic 9th, 10th, 11th, 12th thoracic vertebrae (Fig 1 red arrow) with absent left 9th, 10th, 11th, 12th ribs (Fig 1 blue arrow) and left lumbar hernia (Fig 1 yellow arrow). Ultrasonography abdomen showed a hernial defect in left lumbar region measuring 5 cm X 5 cm containing small bowel loops. Ultrasonography spine showed spina bifida occulta. Echocardiography was normal. Lumbar hernia was repaired by pediatric surgeons, operative findings showed 5 cm X 4 cm hernial sac containing small bowel loops. Child had uneventful post-operative course and discharged successfully. Baby was advised regular follow up.

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Lumbocostovertebral Syndrome (LCVS) is a rare disorder which was first described by Touloukian in 1972.¹ It comprises triad of abnormalities of abdominal wall musculature (Lumbar Hernia), rib and vertebral anomalies. LCVS involves other anomalies like, spinal cord malformations, talipes equino varus deformity, and inguinal hernia etc.^{2,3} It can be associated with VACTERL (*Vertebral, Anal, Cardiac, Trachea-esophageal, Renal, Limb anomalies*) syndrome⁴. Malformations in LCVS occur due to single somatic mutation occurring during 3-5 weeks of gestation³. During this gestational period neurulation occurs hence LCVS involves spinal cord malformations along with other anomalies⁵. Management of LCVS involves thorough evaluation with multidisciplinary approach in correction of malformations and regular follow up.

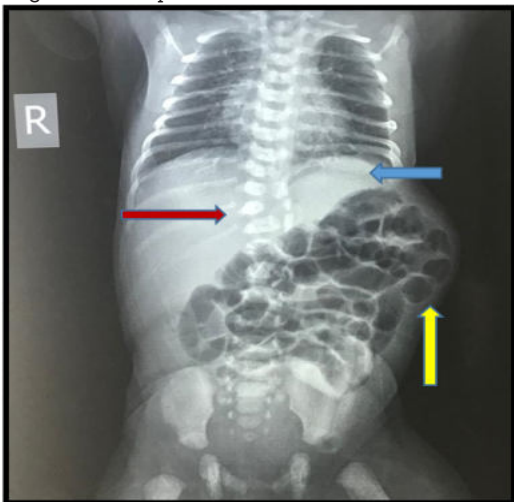


Fig 1 -Roentgenogram showing dysplastic 9th, 10th, 11th, 12th