

# Rare Association of Eventration of Left Hemidiaphragm with Ipsilateral Thyroid Agenesis

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## ABSTRACT

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Eventration is a well-known congenital malformation of the diaphragm, usually asymptomatic and diagnosed incidentally on chest radiography. It is sometimes associated with a number of other congenital syndromes and anomalies. We report a rare case of eventration of left hemidiaphragm associated with gastric volvulus, ipsilateral thyroid agenesis and microphthalmia. [Indian J Chest Dis Allied Sci 2012;54:131-133]

**Key words:** Eventration, Diaphragm, Thyroid gland, Agenesis, Gastric volvulus.

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## INTRODUCTION

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Diaphragmatic eventration is a rare congenital anomaly due to the absence or deficiency of diaphragmatic musculature, radiologically manifested by elevation of the hemidiaphragm.<sup>1</sup> Histopathologically, skeletal muscles of the diaphragm are partially or totally replaced by fibrous tissue. It is usually unilateral, may present at any age, and is more common in male. It can be of two types, partial and complete. Partial eventration is more common on the right side and the complete variety is common on the left side. In adults, unilateral cases are usually asymptomatic; bilateral eventration, although uncommon, may present with respiratory distress. Eventration of the diaphragm may be associated with other congenital anomalies. Here, we report a case of eventration of the left hemidiaphragm associated with organo-axial type of gastric volvulus, ipsilateral microphthalmia, and agenesis of the left hemithyroid. *To the best of our knowledge, ipsilateral thyroid agenesis associated with eventration of the diaphragm has not been reported in the literature.*

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## CASE REPORT

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A 20-year-old, normotensive, non-diabetic man presented with shortness of breath of insidious onset for two years. There was no history of chest pain, haemoptysis, fever or atopy. He had no vision in the left eye since childhood. He was non-smoker and non-alcoholic. There was no history of chest trauma

or surgical intervention on thorax. On interrogation, it was revealed that he was born by a spontaneous uneventful vaginal delivery at 39 weeks of gestation and his birth weight was 2500g. General survey was normal except left-sided microphthalmia (Figure 1). Examination of respiratory system revealed elevation

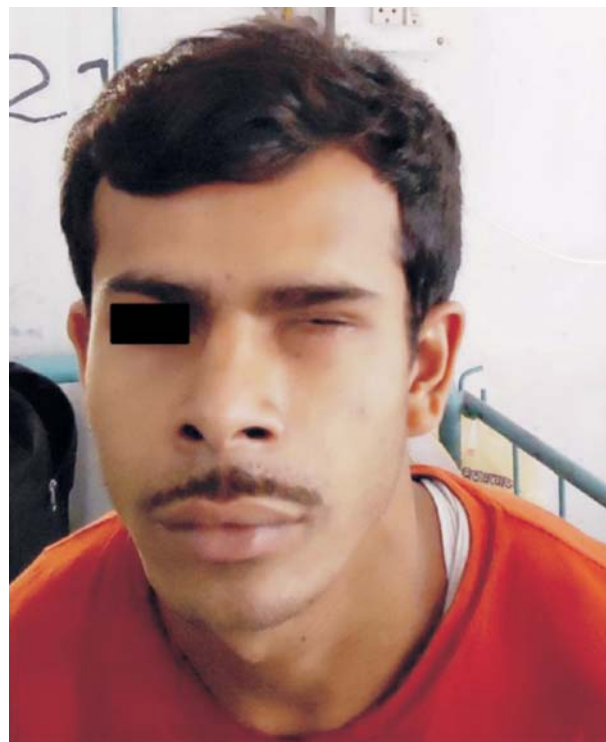


Figure 1. Clinical photograph of the patient showing a left-sided microphthalmia.

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[Received: September 30, 2011; accepted: December 5, 2011]

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of Traube's space, diminished vesicular breath sounds with intermittent peristaltic sounds over left infra-mammary and infra-axillary areas. On ophthalmological examination, right eye was normal with a visual acuity of 6/6, but microphthalmia was detected on left side with no perception of light and projection of rays. Funduscopy of the right eye revealed no abnormality. Examination of other systems was unremarkable.

Complete blood count and blood biochemistry were within normal limits. A chest radiograph showed an elevated left hemidiaphragm with right-sided mediastinal displacement and normal lung parenchyma. This radiological finding was also evident on his old chest radiographs. Ultrasonogram of thorax revealed elevated left dome of the diaphragm with little movement on respiration. On sniffing, paradoxical movement of left hemidiaphragm was noted. Continuity of left hemidiaphragm was preserved and the right hemidiaphragm was normal in all aspects. A contrast-enhanced computed tomography (CECT) of thorax showed eventration of the left hemidiaphragm (Figure 2) and agenesis of the left lobe and the isthmus of thyroid gland (Figure 3). Ultrasonogram of thyroid also confirmed this finding. Thyroid function tests were normal. Barium-meal study revealed a

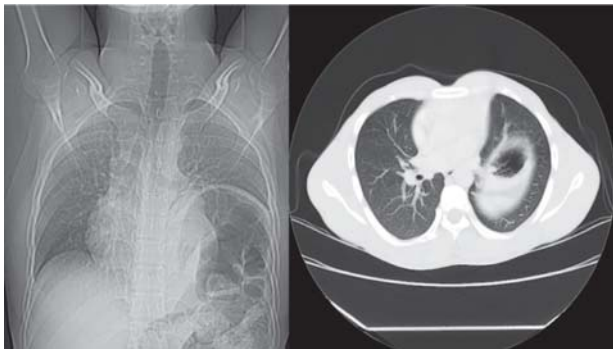


Figure 2. CECT of thorax showing elevated left hemidiaphragm with normal lung parenchyma.



Figure 3. CECT of thorax showing agenesis of left half of thyroid gland and isthmus with intact right half (arrow).

malrotated stomach in the thorax completely on the left side with the greater curvature lying superior to the lesser curvature and duodenal bulb lying to the left, suggestive of organo-axial type of gastric volvulus (Figure 4). An echocardiogram did not reveal any congenital cardiac abnormality. A final



Figure 4. Barium-meal study showing organo-axial type of gastric volvulus.

diagnosis of congenital eventration of the left hemidiaphragm associated with organo-axial type of gastric volvulus, ipsilateral microphthalmia, and agenesis of left hemithyroid was made. The patient was advised for surgical repair, but was lost during follow up.

## DISCUSSION

In eventration of the diaphragm, all or part of it is largely composed of fibrous tissue with a few or no interspersed skeletal muscle fibres, presenting radiologically as elevated hemidiaphragm. It is most commonly congenital but may be acquired. Incidence of congenital eventration is 1 in 10,000.<sup>2</sup> It may be complete or partial. Congenital eventration is most commonly unilateral and is seen on the left side. It is almost always asymptomatic and detected on chest radiographs incidentally. Diaphragmatic movement is reduced, paradoxical or absent on fluoroscopy or on ultrasonogram.<sup>3</sup> Contralateral mediastinal displacement with unilateral elevation of the diaphragm goes more in favour of eventration in contrast to unilateral diaphragmatic palsy, the commonest differential diagnosis of eventration.<sup>4</sup> Eventration allows abdominal contents to enter into the thorax, although the continuity of the diaphragm is intact, in contrast to diaphragmatic hernia.

Congenital eventration of the diaphragm may be associated with other congenital anomalies and syndromes like Poland's syndrome, Beckwith-Wiedemann syndrome, Jarcho-Levin syndrome, congenital rubella syndrome, Kabuki make-up syndrome, gastric volvulus, congenital cytomegalovirus infection, microphthalmia, anophthalmia, congenital heart diseases, pulmonary hypoplasia, tracheomalacia, renal ectopia, deformities of pinna, and Werdnig Hoffman disease, etc.<sup>2</sup> In our patient, left-sided diaphragmatic eventration was associated with microphthalmia, gastric volvulus, and ipsilateral thyroid agenesis.

Because of elevated left hemidiaphragm, wide sub-diaphragmatic area provides the adequate space for malrotation of the stomach, resulting in gastric volvulus.<sup>5</sup> If this rotation occurs along the longitudinal axis connecting the gastro-oesophageal junction with the pylorus, antrum moves from the inferior to superior position. This is called organo-axial volvulus.<sup>6</sup> On the other hand, rotation along the vertical axis extending from the liver to greater curvature results in mesentero-axial volvulus.<sup>6</sup> Association of eventration and gastric volvulus may result in intermittent gastrointestinal symptoms, and sometimes partial or complete gastric outlet obstruction leading to a surgical emergency.<sup>7</sup> Plication of the hemidiaphragm with anterior gastropexy is the surgery-of-choice in this case. In our patient, gastric volvulus was of organo-axial type and fortunately asymptomatic.

After extensive review of the literature, we found that very few cases of diaphragmatic eventration are reported in association with microphthalmia.<sup>8</sup> Congenital structural anomalies of the thyroid gland include hypoplasia or hemiagenesis with or without isthmic agenesis.<sup>9</sup> Prevalence of hemiagenesis is only 0.2 percent.<sup>10</sup> Right-sided hemiagenesis is less common than left one.<sup>11</sup> In most cases of thyroid hemiagenesis, the cause is unknown. It is usually sporadic, but a few familial cases have been reported.<sup>12,13</sup> Patients of congenital thyroid anomalies are euthyroid, although hypothyroidism and

hyperthyroidism may occur.<sup>9</sup> Ultrasonogram is the most useful modality for its diagnosis.<sup>9</sup>

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