CONGENITAL DIAPHRAGMATIC HERNIA

by

S. P. Tyagi,* M.D.

S. K. Dadgar,** M.B.,B.S.

Nirmala Tyagi,*** M.B.,B.S.

Nishat I. Ashraf,† M.D.

and

Kusum Saxena,†† M.S.

Hiatus hernia or diaphragmatic hernia is a very rare congenital anomaly (Harris and Steinberg 1954; Jackson, 1967). Generally there is intrauterine death in these cases but if the baby survives, this constitutes a most urgent neonatal emergency. Most of the infants with acute symptoms die of cardio-respiratory complaints before corrective surgery can be attempted (Koop 1962).

Occasional case reports of congenital diaphragmatic hernia have been published in the Indian Literature (Kelkar *et al* 1959; Paul *et al* 1965; Mathur *et al* 1969; Kumar *et al* 1971 and Vare 1972). The rarity of the disease and associated congenital anomalies have prompted the authors to publish the case.

Case Report

A still born baby was born to a Hindu female having full term pregnancy with breech presentation on 16-12-1976. At birth the baby weighed 2 kg. Heart sounds and respiratory movements were absent. The congenital anomalies noticed were imperforated anus, telipes equinovarus of the right foot and transposition of the scrotum. The parents were persuaded to get the postmortem done so that other congenital abnormalities could be observed.

Post Mortem Report

Body length	— 39 cm.
Circumference of the head	— 30 cm.
Panniculus	— normal
Testes	— undecended
Scrotum	— Ill developed and a small penis like structure was present underneath the scrotum.

On exploring the thoracic cavity it was found that the Left side of the chest cavity was filled with intestine, appendix, liver and spleen and the heart was shifted to the right side (Fig. 1).

Cardio-respiratory System:

Heart weighed 7.2 gms. Both the lungs were very small, particularly the left one which measured 1.0 x 0.9 cm.

Gastro-intestinal System:

Bowels were distended with meconium. Liver weighed 50 gms. and was of dark slaty colour, firm in consistency. Gall bladder and biliary tract were normal.

Urogenital System:

Right kidney weighed 10.6 gms. One big cyst was present in the upper half. Left kidney weighed 14.2 gms. Multiple small cysts were present. Ureters and bladder were normal. Both the testes were undescended, small, bean shaped and measured 0.8 x 0.5 cm.

*Reader in Pathology.

**Research Fellow under Indo-Afghan Cultural Programme.

***Demonstrator in Pathology.

†Lecturer in Pathology.

††Reader in Obstetrics & Gynaecology.

From the departments of Pathology and Obstetrics & Gynaecology, Jawaharlal Nehru Medical College, Aligarh Muslim University, Aligarh.

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Reticuloendothelial System:

Spleen weighed 9.5 gms. Thymus was normal looking.

Microscopic Findings:

Lung, liver and spleen were congested. The morphology of the lungs was that of uninflated organ. Areas of extramedullary haemopoiesis were present in liver, spleen and kidneys.

Sections from the kidneys showed atrophied renal parenchyma, dilated tubular structures separated by hyalinised bands of connective tissue and distended blood vessels.

Discussion

The rarity of congenital diaphragmatic hernia could be visualised by the observations of Harris and Sternberg (1954) and Jackson (1967) who have reported the incidence as 1/8,716 live births and 1/7,000 live births respectively. Hernia can occur either through the foramen of Bochdalek or the foramen of Morgagni or there may be partial or complete absence of diaphragm. In the present case only a thin rim of diaphragm was present at the periphery.

This is more common on the left side resulting into the shifting of the heart to wards the right as had been seen in the case under review. However, diaphragmatic hernia can occur on the right side also but very rarely (Mathur *et al* 1969) and more so it is not usually associated with severe respiratory distress because the liver serves as a barrier (Koop 1962). The proportion of the left sided and right sided hernia varies from 4:1 to 8:1. Hernia through the pleuroperitoneal canal (Foramen of Bochdalek) or through posterio-lateral portion of the diaphragm are generally massive as was observed in the present case and also reported by Harrington (1955).

Though the chest cavity was slightly bulky, more so on the left side and abdomen was flat but the diagnosis of congenital diaphragmatic hernia was never thought of before the postmortem. In living infants the cardinal triad of dyspnoea, cyanosis and dextrocardia may arouse suspicion of the possibility of congenital diaphragmatic hernia.

Other congenital abnormalities may be associated with congenital diaphragmatic hernia like anencephaly, spina bifida, meningocele (Vare 1972) or at times agenesis of the lung (Chaireaux and Ferreiara 1958; Kumar *et al* 1971) but polycystic kidneys, undescended testes, transposition of the scrotum and imperforated anus were a rare combination and that had caused an interest among the authors to publish the case.

Summary

A postmortem report of a still born baby having congenital diaphragmatic hernia has been reported. Besides hernia, other congenital abnormalities present were imperforated anus, transposition of scrotum, undecended testes, telepes equinovarus and polycystic kidneys.

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See Fig. on Art Paper V