J. Fetal Med. (September 2017) 4:153–155
DOI 10.1007/s40556-017-0131-5

BRIEF COMMUNICATION

Antenatal Diagnosis of Congenital Diaphragmatic Hernia and Successful Outcome After Postnatal Surgery

Deval Shah[1] · P. K. Vaghela[2] · Suresh Patel[3] · Prashant Acharya[4]

Received: 24 May 2017 / Accepted: 5 July 2017 / Published online: 29 July 2017

Abstract Congenital diaphragmatic hernia (CDH) is a group of disorders where abdominal contents protrude into the chest. The prognosis depends on the contralateral lung volume. The authors evaluated a case of left sided diaphragmatic hernia that was diagnosed at 32 week, and confirmed by MRI. Sonography based Lung to Head circumference ratio (LHR) was calculated. The case was managed at a tertiary care centre where the newborn was immediately shifted to the pediatrician and pediatric surgeon and was operated. Antenatal sonography is the primary modality to diagnose CDH. LHR based on sonography is the key to predict pulmonary hypoplasia and, an MRI is additive. The management needs a multidisciplinary approach.

Keywords Congenital diaphragmatic hernia · Four chamber view · Contralateral lung volume · Lung to head circumference ratio

✉ Deval Shah
devalnilpa@gmail.com

[1] Department of Gynaecology, Astha Hospital and Fetal Medicine Centre, Deesa, Gujarat, India

[2] Department of Gynaecology, Satyam Hospital, Deesa, Gujarat, India

[3] Department of Radiology, Banas Sonography Centre, Deesa, Gujarat, India

[4] Department of Fetal Medicine, Paras Fetal Medicine Centre, Ahmedabad, Gujarat, India

Introduction

Congenital diaphragmatic hernia is a group of disorders in which part of abdominal contents protrude into the chest. The incidence is 1 in 2000–3800 [1], in isolated cases, the prognosis depends on contralateral lung volume estimated by sonography and MRI may add value to prognosticate.

Case Report

Mrs. ABC, a 32-year-old lady having active married life of 6 year who was 2nd gravida with one previous abortion was referred for growth scan at 7 month amenorrhea. She had no complaints and a previous anomaly scan done at 22 week at authors' hospital showed no obvious anomaly on four chamber view (Fig. 1). Her nuchal translucency (NT) scan was normally reported. The ultrasound showed normal growth and corresponded to 32 week; the liquor was adequate and doppler study was normal. Stomach was seen in the abdomen at its normal position (Fig. 1). Examination of the fetal chest at 4 chamber view revealed congenital diaphragmatic hernia (Fig. 1).

Course of the superior mesenteric artery was upturned towards the chest (Fig. 1). For antenatal prediction of postnatal prognosis, contralateral lung volume was measured by trace method and observed to expected (O/E) Lung to Head circumference ratio (LHR) was calculated which was found to be 84%.

Fetal MRI was done and that revealed a 23 mm defect on the left posterolateral diaphragm with small bowel in the chest and the heart was shifted to the right (Fig. 2). No other associated anomalies were found. The fetal karyotype was normal.

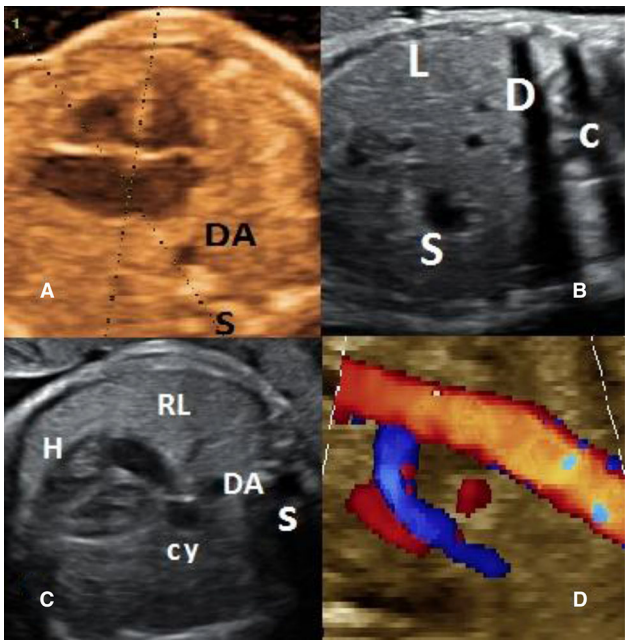

Fig. 1 Antenatal ultrasound **a** Four chamber view (axial section) at 22 week, **b** stomach seen at normal position at 32 week, **c** four chamber view at 32 week, **d** upturn course of superior mesenteric artery at 32 week. *C* Chest; *cy* Cyst; *D* Diaphragm; *DA* Descending aorta; *H* Heart; *L* Liver; *Rl* Right lung; *S* Spine

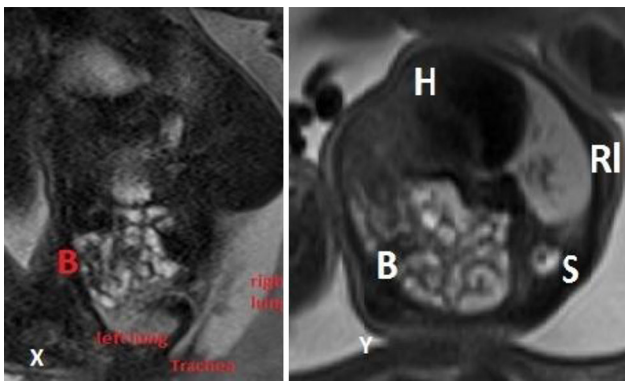

Fig. 2 MRI at 33 week. *X* Coronal image; *Y* Transverse image at four chamber heart. *B* Bowel; *H* Heart; *Rl* Right lung; *S* Spine

Spontaneous onset of labour at 35 week occurred and a 2.0 kg male child was delivered vaginally. The neonate was admitted to the neonatal intensive care unit (NICU). His spO_2 was 90%. He was shifted to the pediatric surgeon with ventilator support after 48 h and was operated. There was a posterolateral left sided defect (BOCKDALEK type) of around 22–24 mm. The intestines were reduced to the abdomen and the closure was done by vicryl 3.0. There was intestinal malrotation which was corrected by Ladd's procedure. Appendicectomy was done. Immediately after the surgery, the child was admitted back to the NICU where he was kept for 11 day and the further postnatal course was uneventful.

Discussion

CDH is a malformation of the diaphragm due to failure of closure of pleuroperitoneal canal at around 10 week leading to herniation of abdominal contents into the thoracic cavity leading to compression of lung tissues at pseudo glandular stage. Early prenatal diagnosis is associated with 2–4 times increase in postnatal mortality which may be due to large defect and associated anomalies. Mean age of diagnosis is around 24 week. About 50% cases are isolated and the survival depends on the contralateral lung volume. Around 25% cases are associated with chromosomal anomalies and 25% with other major anomalies or some syndrome [1].

Ninety percent cases are due to posterolateral defect which is also called Bochdalek hernia—80% are left sided, 15% are right sided and 5% are bilateral. Left sided defect leads to protrusion of stomach, bowels, spleen and left liver lobe into the left chest along with right sided mediastinal shift with dextroposition of the heart. Herniated viscera leads to pulmonary hypoplasia and pulmonary hypertension [2].

Prognosis depends on the gestational age at diagnosis (earlier the diagnosis, likely larger the defect), associated anomalies, contralateral lung volume, pulmonary vasculature (Doppler), cardiac function, liver herniation and gestational age at delivery.

Although majority of cases are sporadic, diaphragmatic hernia can also be a part of many syndromes (Table 1). Lung to head circumference ration (LHR) is indirect 2D assessment of pulmonary hypoplasia [3]. It is the measurement of the contralateral lung in 4 chamber view of heart by trace or diameter method. Tracing method is better in predicting the lung volume. Head circumference is measured by Elliptical method in mm.

$$\text{Lung to head ration (LHR)} = \text{lung area}/\text{head circumference}.$$

If LHR is 1 or less—the prognosis is poor, if LHR is between 1 and 1.4—extracorporeal membranous oxygenation (ECMO) is often required and if LHR is more than 1.4—the prognosis is good.

In healthy fetus, LHR increases with gestational age, and this can be overcome by Observed to Expected LHR which is independent of the gestational age. Observed LHR is expressed as percentage of Expected mean for gestational age (GA).

$$\text{O/E LHR} = (\text{Observed LHR}/\text{Expected LHR}) \times 100$$

Survival is nearly 100% with O/E LHR > 45%, 50% with LHR 25–45% and poor with LHR < 25%.

Fetal MR imaging is valuable for anatomic assessment [3, 4], determining the organ herniated particularly the liver

Table 1 Syndromes associated with congenital diaphragmatic hernia

Syndrome	Brief description
Fryns syndrome	CDH, coarse facial features, cleft lip/palate, heart disease, cerebral malformations, finger hypoplasia
Thoraco-abdominal syndrome	Ventral hernia, hypoplastic lungs, cardiac anomalies
Beckwith–Wiedemann syndrome	Visceromegaly, macroglossia, anterior abdominal wall defects
CHARGE syndrome	Coloboma, choanal atresia, growth retardation, cardiac, genital and ear abnormalities
Simpson–Golabi–Behmel syndrome	Macrosomia, coarse facial features, macroglossia, skeletal and renal abnormality, abdominal wall defect
Wolf–Hirschhorn syndrome	"Greek helmet" facial appearance, mental/growth retardation
Cornelia de Lange syndrome	Abnormal facial features, growth retardation, microcephaly, upper limb defect
Pallister–Killian syndrome	Coarse facial features with broad forehead and hypertelorism, sparse temporal hair line, hypopigmentation, mental retardation

CDH congenital diaphragmatic hernia

and the effect of the herniated contents on adjacent structures, associated malformations and calculation of the diameters of the pulmonary arteries and aorta. It is helpful in counseling and planning. Based on LHR calculated by MRI, prognostic value for survival for left sided CDH is superior to sonography based LHR as MRI is more accurate in measurement of lung volume than sonography. MRI will not replace sonography as the 1st line screening technique for CDH. However, particularly when sonographic imaging conditions are impaired (oligohydramnios, maternal obesity, unfavorable fetal position), fetal MRI is a feasible technique.

Conclusions

In the last decade, there has been an increase in survival rate of CDH mainly due to prenatal diagnosis, better surgical techniques and ventilator support. Sonography is the primary modality for the diagnosis and LHR based on sonography is the key to predict pulmonary hypoplasia in isolated cases and for patient counseling. MRI is additive to the diagnosis. The management needs multidisciplinary approach.

Acknowledgements Dr Prashant Acharya, Paras Fetal Medicine Centre, Ahmedabad; Radiscan Imaging Centre, Ahmedabad; Dr Amar A Shah, Pediatric Surgeon, Amardeep Hospital, Ahmedabad; Dr Ashish Mehta, Arpan Newborn Care Centre, Ahmedabad.

Compliance with Ethical Standards

Conflict of Interest None.

Source of Funding None.

References

1. Centers for disease control and prevention: birth defects: data and statistics. Available at: http://www.cdc.gov/ncddd/birthdefects/data, 2015. Accessed in March 2017.
2. Richards DS, Kays DM. Fetal umbilical vein deviation in congenital diaphragmatic hernia. J Ultrasound Med. 2013;32:263–8.
3. Kehl S, Siemer J, Brunnemer S, et al. Prediction of postnatal outcomes in fetuses with isolated congenital diaphragmatic hernias using different lung to head circumferance ratio measurements. J Ultrasound Med. 2014;33:759–69.
4. Bebbingtom M, Victoria T, Danzer E, et al. Comparison of ultrasound and magnetic resonance imaging parameters in predicting survival in isolated left-sided congenital diaphragmatic hernia. Ultrasound Obstet Gynecol. 2014;43:670–4.