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Congenital diaphragmatic hernia with intrathoracic testis: A rare entity

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Abstract
Congenital diaphragmatic hernia (CDH) is a major congenital malformation where abdominal contents herniate into the chest. The usual contents of left-sided CDH are the small and large bowels, the stomach, the spleen, the left lobe of liver, and occasionally, the kidney. It is rare for the testis to be part of the herniated contents as compared to other intra-abdominal or retroperitoneal organs. We present a case of intrathoracic testi's occurring in a Bochdalek's CDH managed at our centre.

Keywords: Congenital diaphragmatic hernia, Testis

Introduction
Congenital diaphragmatic hernia is a developmental defect of the diaphragm causing herniation of abdominal contents into thoracic cavity. It occurs in approximately 1 in 2500 to 5000 children and results in high mortality and morbidity [2]. The content of the hernia is usually intra-abdominal or retroperitoneal organs. There have been case reports of testes in the thoracic cavity, however, these are rare [3, 4]. Most of the cases were CDH with intrathoracic unilateral testis. Here, we present the first reported case of left posterolateral congenital diaphragmatic hernia (Bochdalek type) with intrathoracic contralateral testis.

Case Report
A full-term male baby at 39 weeks of gestation was born per-vaginal with birth weight of 2,530 grams and was diagnosed with left-sided CDH. The child was subsequently intubated in view of worsening respiratory distress at 10 minutes of life. Physical examination showed hyper inflated chest with non-palpable right testis in scrotum. Chest radiograph revealed left sided CDH. We proceeded with CDH repair on day 6 of life via an abdominal approach. The hernia contents were large bowel, small bowel, stomach, spleen, pancreas, right testis, and part of right kidney as showed in Figure 1(A). Even though right testis was found intrathoracically, we did not proceed to fix the right testes. Postoperatively, the child recovered well and remains under follow up. We are planning for a laparoscopic examination later.

Discussion
The pathogenesis of CDH remains unclear. Generally, development of the diaphragm occurs from 4th to 12th weeks of gestation. It is derived from fusion of septum transversum, the pleuroperitoneal folds (PPFs), oesophageal mesentery, and body wall mesoderm. It was initially thought that CDH is due to the failure of closure of pleuroperitoneal canal during 8th to 10th weeks of gestation [3, 5]. This causes abdominal organs to herniate into the chest during the time of lung development. The herniation leads to fetal abnormal breathing movement and results in lung immaturity. Subsequently, leading to pulmonary hypoplasia. Another theory is that lung hypoplasia is the causal factor of CDH. Impaired development of post hepatic mesenchymal plate (PHMP) which is closely related to lung development may lead to defective diaphragm [6]. Dual-hit hypothesis explains pulmonary hypoplasia in nitrofen-induced rat model [7, 8]. Initial insult occurs during organogenesis which impairs lung's development caused by nitrofen. Second hit is compression of ipsilateral lung by the herniated abdominal contents. Recent cytogenetic analysis correlates COUP-TFII with CDH formation [5, 7, 8]. COUP-TFII is essential in formation of diaphragm which expressed in primitive foregut mesenchyme, the developing PHMP, developing lung, and septum transversum. Its ablation, hence causing CDH formation.

Other perspective includes disruption of retinoid-signaling pathway in nitrofen model of CDH which causes pulmonary hypoplasia. Thus, it is being practiced for retinoic acid to be started in prenatal period for prevention of lung hypoplasia even though it is controversial.

The most common type of CDH is posterolateral or known as Bochdalek hernia that accounts for 70 - 75% of cases. Majority would be on the left side (85%) [4]. It is rare for the testes to be among the herniated contents. Undescended testis (UDT) or known as cryptorchidism is the failure of one or both testes to descend into the scrotum. It affects in 3% of full-term male baby with 30% in premature ones [9, 10]. The proximity to pleuroperitoneal fold together with shared developmental processes provide an evidence in association between CDH and UDT [3]. The process of testicular descent involves two phases: transabdominal and inguinoscrotal phases. In initial phase, it takes place at 8 - 15 weeks of gestation. It involves enlargement of gubernaculum and regression of cranio-suspensory ligament (CSL) which are under control of Mullerian-inhibiting substance (MIS) and insulin-like hormone 3 (Insl3) [11]. Mutation in both hormones impede growth of gubernaculum that will hinder the testes into internal inguinal ring. Second stage occurs in between 25 - 35 weeks of gestation as gubernaculum and testis migrate from inguinal canal to the scrotum. This phenomenon is under control of genitofemoral nerve with its sensory neuropeptide, calcitonin gene related peptide (CGRP) which help to guide the migration [11]. Defect in the diaphragm leads to abnormality of CSL and disrupt testicular descent. Hypertrophy of gubernaculum causes testis to be migrated into the thoracic cavity on the other hand.

The primary treatment for undescended testis is surgical repositioning of testis into the scrotum or known as orchidopexy. In case of UDT in CDH, orchidopexy is usually performed a couple years later as in work of Hisano *et al*. by laparoscopic technique [12]. This method is able to localize position of testis accurately and safely and minimizes complication in postoperative patients with CDH. We support a laparoscopic approach due to these reasons. We caution against performing an orchidopexy during the initial surgery to avoid prolonged anaesthesia and higher risk of damage to the neonatal testis during fixation.

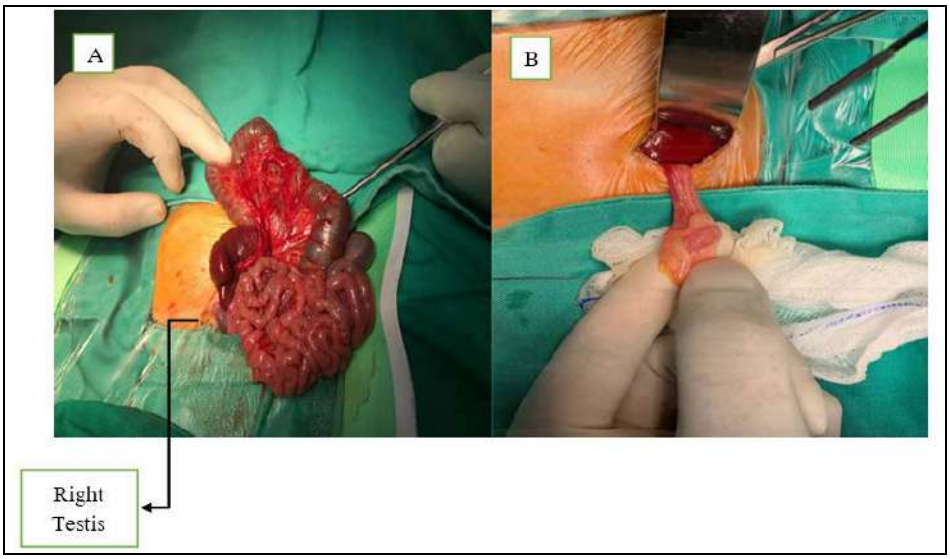

Fig 1: A) Right testis found in conjunction with other abdominal organs through left upper transverse incision B) Right testis brought out with large gubernaculum

Conclusion

In summary, intrathoracic testis in confluence with congenital diaphragmatic hernia remains as a rare entity especially involving contralateral testis. Basic principles of intra- abdominal testes fixation will follow post replacement of the testes into the abdominal cavity. Laparoscopic assessment after one year of age would be an option in these children.

References

1. Tovar JA. Congenital diaphragmatic hernia. Orphanet Journal of Rare Diseases 2012;7:1. doi: 10.1186/1750-1172-7-1.
2. Matthew Clifton S, Mark Wulkan L. Congenital diaphragmatic hernia and diaphragmatic eventration. Clinics in perinatology 2017;44(4):773-779.
3. Azarow KS, Cusick R, Wynn J, *et al*. The association between congenital diaphragmatic hernia and undescended testes. Journal of Pediatric Surgery 2015;50(5):744-745.
4. Mehran Hiradfar, Sadegh Sadeghipoor. Intrathoracic testicular ectopia in congenital diaphragmatic hernia. Asian Journal of Surgery 2006;29(4):303-5.
5. Maria Arafah, Deena Boqari T, Khaled Alsaad O. Left-sided congenital diaphragmatic hernia with multiple congenital cardiac anomalies, Hernia Sac, and Microscopic hepatic heterotopia: A Case Report. SAGE-Hindawi Access to Research Pathology Research International 2011, Article ID 967107, 4 pages. Doi: 10.4061/2011/967107.
6. Chandrasekharan *et al*. Maternal Health, Neonatology, and Perinatology 2017;3:6. doi: 10.1186/s40748-017-0045-1.
7. Keijzer R, Liu J, Deimling J, Tibboel D, Post M. Dual hit hypothesis explains pulmonary hypoplasia in the nitrofen model of congenital diaphragmatic hernia. The American Journal of Pathology 2000;156:1299-306.
8. Puri P, Zimmer J. Congenital diaphragmatic hernia. Newborn Surgery. 4th Ed. Dublin, Ireland: CRC Press 2018.

9. Klonisch T, Fowler PA, Hombach-Klonisch S. Molecular and genetic regulation of testis descent and external genitalia development. Developmental Biology. 2004;270(1):1-18. Doi: 10.1016/j.ydbio.2004.02.018.
10. Husmann DA, Levy JB. Current concepts in the pathophysiology of testicular undescent. Urology 1995;46(2):267-76. Doi: 10.1016/s0090-4295(99)80207-6.
11. Hutson JM, Balic A, Nation T, Southwell B. Cryptorchidism. Seminars of Pediatric Surgery 2010;19(3):215-24. Doi: 10.1053/j.sempedsurg.2010.04.001.
12. Hisano K, Kurisu S, Okada M. Laparoscopic orchiopexy of intraabdominal undescended testis associated with congenital diaphragmatic hernia. Surgery Today 1998;28(11):1224-6. doi: 10.1007/s005950050321.