

Congenital Bronchobiliary Fistula in Association With Right-Sided Congenital Diaphragmatic Hernia

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A case of congenital bronchobiliary fistula is reported in conjunction with another congenital anomaly, right-sided congenital diaphragmatic hernia. The liver was herniated through the diaphragmatic defect into the right chest where a fistula tract was identified from the surface of the liver to the right mainstem bronchus. The patient was treated successfully.

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INDEX WORDS: Congenital bronchobiliary fistula, congenital tracheobiliary fistula, congenital diaphragmatic hernia.

CONGENITAL bronchobiliary fistula is an extremely rare anomaly with a small number of cases reported in the literature.¹⁻¹³ Only 2 previous cases were associated with other congenital malformations.^{8,13} Here we report on a newborn infant with right-sided congenital diaphragmatic hernia (CDH) who had a bronchobiliary fistula from the right mainstem bronchus to the left lobe of the liver discovered at surgery.

CASE REPORT

The baby was 37-week-gestation boy weighing 1,700 g who was born in severe respiratory distress. CDH had not been diagnosed antenatally. After intubation in the delivery room, chest x-ray showed a right-sided CDH. The patient was explored later through a right subcostal incision. There was a large diaphragmatic defect with a hernia sac present. Multiple loops of small bowel and colon were present in the right chest. The left lateral segment of the liver was intraabdominal, but the remaining lobes were above the diaphragmatic defect. There was significant difficulty mobilizing the liver out of the right chest. Dissection showed a distinct connection between the cephalad surface of the liver and the inferior aspect of the right lung near the hilum. Further dissection found a 4-mm-diameter x 4-mm-long tubular structure connecting the surface of the left medial lobe of the liver to the right mainstem bronchus (Fig 1). When divided, the cephalad portion had the appearance of bronchial tissue, with cartilaginous rings and air leaking from it in concert with ventilation. The caudad portion entered the surface of the left lobe of the liver and was draining bile. Both defects were closed primarily. The patient survived. The fistula was not of sufficient length to yield a surgical specimen, and patient instability precluded further intraoperative diagnostic studies.

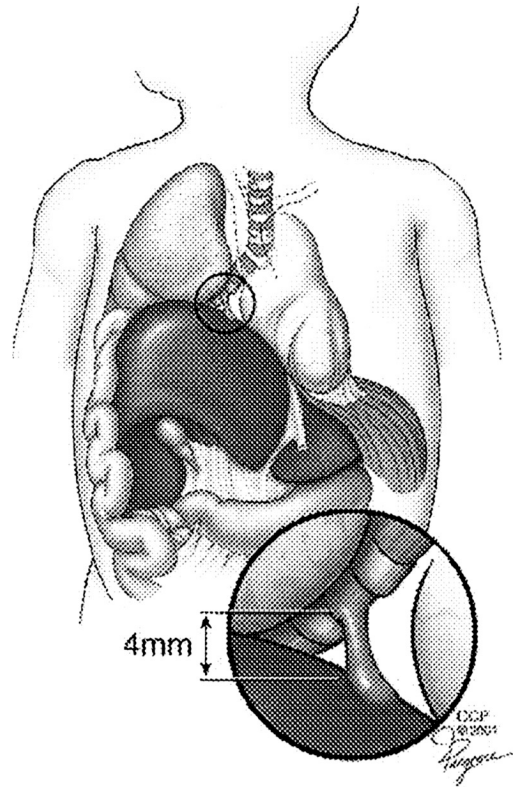


Fig 1. Artist's illustration of operative findings: bronchobiliary fistula from left lobe of liver to right mainstem bronchus. Reprinted with the permission of Cleveland Clinic Foundation.

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DISCUSSION

To date, there are only several reported cases of congenital bronchobiliary fistula, the majority of which were treated successfully.¹⁻¹³ Most patients presented with chronic respiratory symptoms and recurrent pneu-

monia, usually starting in early infancy. Patient ages ranged from several days to several years; however, there are reports of bronchobiliary fistula in adults.^{14,15} In most cases, a fistula tract several centimeters in length was found originating from the region of the carina, passing through the esophageal hiatus, and entering the left lobe of the liver, ultimately communicating with the left hepatic duct. In all but one,⁶ the bronchial opening was to the right side of the carina or to the right mainstem bronchus, as in the current case. Most were diagnosed preoperatively by bronchoscopy and associated radiographic contrast studies through the fistula. There is only one other report of bronchobiliary fistula found incidentally at surgery—a case in which the anomaly was encountered during repair of esophageal atresia/tracheo-esophageal fistula.⁸ Histology of resected specimens generally shows bronchial tissue proximally with characteristics of esophagus or bile duct distally.^{2,3,5}

This is the third case of congenital bronchobiliary fistula reported in association with another congenital anomaly.^{8,13} In our case, the bronchobiliary fistula was an unexpected operative finding. We were unable to show communication of the fistula with the hepatic ductal system by contrast radiography. However, after division of the fistula, the observations of air leakage from the proximal portion of the fistula and the appearance of bile in the distal portion of the fistula support the diagnosis.

Two hypotheses concerning this anomaly have been proposed. One suggests this anomaly results from the joining of an accessory bronchus with an aberrant hepatic duct.¹⁶ The other suggests the fistula represents a duplication of the upper gastrointestinal tract.¹ Embryologically, the fact that the fetal lung originates as a ventral diverticulum of the pleuripotential foregut^{17,18} makes an analogous origin for respiratory and enteric malformations seem plausible.

REFERENCES

1. Neuhauser EB, Elkin M, Landing B: Congenital direct communication of the biliary system and the respiratory tract. *Am J Dis Child* 83:654, 1952
2. Enjoji M, Watanabe H, Nakamura Y: A case report: Congenital bilitracheal fistula with trifurcation of bronchi. *Ann Paediatr* 200:321-332, 1963
3. Stigol LC, Traversaro J, Trigo ER: Carinal trifurcation with congenital tracheobiliary fistula. *Pediatrics* 37:89-91, 1966
4. Haight C, Graves WH: As quoted by Weitzman JJ, Cohen SR, Woods L et al: Congenital bronchobiliary fistula. *J Pediatr* 73:329-334, 1968
5. Weitzman JJ, Cohen SR, Woods L, et al: Congenital bronchobiliary fistula. *J Pediatr* 73:329-334, 1968
6. Wagget J, Stool S, Bishop H, et al: Congenital broncho-biliary fistula. *J Pediatr Surg* 5:566-569, 1970
7. Sane SM, Sieber WK, Girdany BR: Congenital bronchobiliary fistula. *Surgery* 69:599-608, 1971
8. Kalayoglu M, Olcay I: Congenital bronchobiliary fistula associated with esophageal atresia and tracheo-esophageal fistula. *J Pediatr Surg* 11:463-464, 1976
9. Dyon JF, de Marliave H, Bacle B, et al: Les fistules aerodigestives congenitales rare. *Chir Pediatr* 25:234-244, 1984
10. Lindahl H, Nyman R: Congenital bronchobiliary fistula successfully treated at the age of three days. *J Pediatr Surg* 21:734-735, 1986
11. Egrari S, Krishnamoorthy M, Yee CA, et al: Congenital bronchobiliary fistula: Diagnosis and postoperative surveillance with HIDA scan. *J Pediatr Surg* 31:785-786, 1996
12. Gauderer MW, Oiticica C, Bishop HC: Congenital bronchobiliary fistula: Management of the involved hepatic segment. *J Pediatr Surg* 28:452-455, 1993
13. Chan YT, Ng WD, Mak WP, et al: Congenital bronchobiliary fistula associated with biliary atresia. *Br J Surg* 71:240-241, 1984
14. Yamaguchi M, Kanamori K, Fujimura M, et al: Congenital bronchobiliary fistula in adults. *South Med J* 83:851-852, 1990
15. De Carvalho CR, Barbas CS, Guarnieri RM, et al: Congenital bronchobiliary fistula: First case in an adult. *Thorax* 43:792-793, 1988
16. Bremer JL: Congenital anomalies of the viscera: Their embryological basis. Cambridge, MA, Harvard University Press, 1957
17. Boyden EA: Development and growth of the airways, in Hodson WA (ed): *Development of the Lung*. New York, NY, Dekker, 1977
18. DiFiore JW, Wilson JM: Lung development. *Semin Pediatr Surg* 3:221-232, 1994