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Research Letter

Treatment of fetal congenital chylothorax: Report of eight cases at a mainland Chinese medical center

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Dear Editor,

Congenital chylothorax, defined as the accumulation of chyle in the pleural space, occurs after disruption of the thoracic duct. Although a rare neonatal disorder, it is the most common cause of pleural effusion in fetuses and newborns. The incidence of congenital chylothorax is reported as 1 in 2000 NICU admissions [1]. Chylothorax produces detrimental respiratory, nutritional, and immunological consequences. Mortality rates range from 20% to 60% depending upon associated findings, gestational age, and the duration and severity of the chylothorax [2]. It is thus important to develop methods that can decrease the morbidity and reduce hospital stay. However, management practices vary among units and no consensus has been reached. Here we first report on the experience with the management of congenital chylothorax at a mainland Chinese medical center, resulting in good outcomes.

From January 2014 to December 2015, we had eight cases (six males, two females) of congenital chylothorax admitted to the surgical neonatal intensive care unit (SNICU) at the Guangzhou Women and Children's Medical Center, which is a tertiary perinatal and neonatal unit in southern China (Table 1). Prenatal diagnosis was made in all patients. The diagnosis of pleural effusion was made prenatally via ultrasound. Hydrops fetalis was defined as a fetus with two or more sonographic findings of excess fluid accumulated in the form of ascites, pleural or pericardial effusions, skin edema, placental edema, or polyhydramnios. The chylothorax was confirmed based on the criteria with triglyceride levels of >110 mg/dL, or a total white blood cell count >1000/μL with a predominance of lymphocytes [3]. Chromosomal abnormality was excluded using chromosomal microarray analysis of cells in the pleural effusion.

In our practice, gestational age and the presence of hydrops were the two important factors in deciding treatment of congenital chylothorax, Because thoraco-amniotic shunting was not available at our center, we only proceeded conservatively at first with close follow-up when the gestational age was less than 32 weeks. Prenatal thoracocentesis might be used if necessary. When the gestational age reached 32 weeks, prompt delivery was offered if the pleural effusion progressed quickly or hydrops developed. Thoracocentesis prior to delivery was performed to facilitate neonatal resuscitation.

After birth, postnatal management of chylothorax started at the labor ward. A neonatologist and a neonatal surgeon accompanied the birth. Neonatal pulmonary function was evaluated promptly and accurately. If respiratory distress occurred, intubation was provided. Closed-chest drainage was established to remove fluid from the pleural space. Other treatment options were mechanical ventilation, replacement of albumin and globulin loss, prevention of infections, and dietary modification.

The mean gestational age at diagnosis was 31.7 (22.6–38) weeks, and the median gestational age at birth was 35.3 (33.3–39.6) weeks. There were two full-term cases and six premature cases. The birth weight ranged from 2320 to 3720 g. Seven cases survived, and none needed surgical treatment. The median duration of mechanical ventilation was 24 days (7–57 days). The median time course of resolution for pleural effusion was 23 days (15–47 days). The median hospitalization time was 26 days (18–72 days).

Two cases had unilateral (right-sided) pleural effusion, and presented at full-term gestation. They needed no mechanical ventilation, and had a shorter duration of pleural effusion resolution and hospital stay. Six cases had bilateral pleural effusion with a premature delivery, and all required the mechanical ventilation. Case 3 had prolonged ventilation and hospitalization durations because of repeated chest tube falling out and placements. Case 1 first presented with bilateral pleural effusion at 24 weeks'

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Table 1
Congenital chylothorax: perinatal, neonatal details and associated anomalies.

Case	Sex	GA at diagnosis (weeks)	GA at delivery (weeks)	Prenatal thoracocentesis	Location	Hydrops	Associated anomalies	Delivery mode	Birth weight (g)	Apgar score (1/5 min)	Intubation/drainage at birth	MV duration (d)	Resolution duration (d)	Hospitalization duration (d)	Outcome
1	M	24.1	33.3	Nil	Bilateral	Present	–	CS	2350	6/8	Yes/Yes	21	–	–	Died
2	M	22.6	33.4	Yes	Bilateral	Present	–	CS	3200	3/5	Yes/Yes	26	34	39	Alive
3	M	33.3	34.7	Yes	Bilateral	Nil	–	VD	2490	8/9	No/Yes	58	47	72	Alive
4	F	35.1	36.1	Yes	Bilateral	Nil	VSD	VD	2320	8/9	No/Yes	7	15	22	Alive
5	F	26	36	Yes	Bilateral	Present	–	CS	3700	4/8	Yes/Yes	22	18	30	Alive
6	M	30	34.1	Yes	Bilateral	Present	–	CS	3720	5/7	Yes/Yes	40	23	46	Alive
7	M	38	39.6	Nil	Right	Nil	–	VD	3120	8/9	No/Yes	–	14	22	Alive
8	M	37.4	38.6	Nil	Right	Nil	–	VD	2920	1/5	Yes/Yes	–	5	18	Alive

CS, caesarean section; *GA*, gestational age; F, female; M, male; MV, mechanical ventilation; VD, vaginal delivery; VSD, ventricular septal *defect*.

gestation. However, the mother did not come back until 33 weeks when she had the signs of premature labor. A repeat scan found fetal bilateral pleural effusion with hydrops fetalis. An emergency cesarean was followed, and the newborn was intubated and under high frequency ventilation and closed-chest drainage. Unfortunately, a chest X ray revealed bilateral pneumothoraces (Fig. 1). The infant died of respiratory failure 21 h after birth.

In this report, we describe our experience of management of fetal congenital chylothorax at a single mainland Chinese perinatal center. The overall 87.5% of survival rate is higher than that (71%) reported in a recent study [4]; the total length of hospital stay (18–72 days) is also shorter than that (29–145 days) reported in another study [5]. These differences might be due to the small sample size in our serials. Although the optional management of fetal congenital chylothorax in the prenatal period is still a matter of debate, from our experience in treating these cases, we believe that early diagnosis, prenatal thoracocentesis, and aggressive initiation of non-operative postnatal management options can decrease the mortality rate of this disorder. The benefits of our strategy were assumed to be the potential effects in the resolution of pleural effusions and the prevention of pulmonary hypoplasia by antenatal intervention, and the facilitation of neonatal resuscitation and respiratory management in the delivery room. A recent study also found that infants who subsequently received prenatal therapy experienced a better perinatal condition and exhibited improved postnatal outcomes compared to infants who did not receive prenatal therapy [4]. However, preterm birth <34 weeks and low birth weight are two independent adverse prognostic factors in fetal congenital chylothorax [6,7]. Therefore the timing of delivery in such cases must balance the newborn risks of late-preterm and early-term delivery with the risks of fetal hydrops. Decisions regarding timing of delivery should be individualized. Due to the fact that congenital chylothorax is a life-threatening condition, reporting such cases can be a great assistance to physicians for managing the disease, and also help with defining the ideal algorithm for guiding the management in the prenatal and neonatal

Fig. 1. Chest X-ray (AP view) showing bilateral pneumothoraces at 4 h of life.

periods. Indeed, we are trying to establish the thoracoamniotic shunting in fetuses with early-onset massive pleural effusion at our center, since fetal chest shunt insertion seems to have a better outcome [8,9].

Conflicts of interest

The authors have no conflicts of interest relevant to this article.

References

[1] Van Aerde J, Campbell AN, Smyth JA, Lloyd D, Bryan MH. Spontaneous chylothorax in newborns. Am J Dis Child 1984;138:961–4.
[2] Dubin PJ, Kind IN, Gallagher PG. Congenital chylothorax. Curr Opin Pediatr 2000;12:505–9.
[3] Buttiker V, Fanconi S, Burger R. Chylothorax in children: guidelines for diagnosis and management. Chest 1999;116:682–7.
[4] Lee CJ, Tsao PN, Chen CY, Hsieh WS, Liou JY, Chou HC. Prenatal therapy improves the survival of premature infants with congenital chylothorax. Pediatr Neonatol 2016;57:127–32.
[5] Downie L, Sasi A, Malhotra A. Congenital chylothorax: associations and neonatal outcomes. J Paediatr Child Health 2014;50:234–8.
[6] Bengtsson BO. Outcome of neonatal chylous effusions: what do you tell the parents? A 10-year experience in a community hospital. Am J Perinatol 2014;31:1037–42.
[7] Caserío S, Gallego C, Martin P, Moral MT, Pallás CR, Galindo A. Congenital chylothorax: from foetal life to adolescence. Acta Paediatr 2010;99:1571–7.
[8] Yinon Y, Grisaru-Granovsky S, Chaddha V, Windrim R, Seaward PG, Kelly EN, et al. Perinatal outcome following fetal chest shunt insertion for pleural effusion. Ultrasound Obstet Gynecol 2010;36:58–64.
[9] Jeong BD, Won HS, Lee MY, Shim JY, Lee PR, Kim A. Perinatal outcomes of fetal pleural effusion following thoracoamniotic shunting. Prenat Diagn 2015;35:1365–70.