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Microcystic congenital pulmonary airway malformation with hydrops fetalis: steroids vs open fetal resection

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Abstract

Background/Purpose: Congenital pulmonary airway malformations (CPAM) are rare lesions often diagnosed during routine prenatal ultrasound. The presence of hydrops fetalis is an indicator of poor prognosis. Here we present a retrospective review of fetuses undergoing either open fetal surgery or steroids for predominantly microcystic CPAM with hydrops fetalis.

Method: A retrospective review of patients undergoing open fetal surgery or steroids for CPAM at our institution was performed. The primary outcome was survival.

Results: A retrospective review of all patients referred to our institution with the diagnosis of CPAM was performed. Fetuses with predominantly microcystic CPAM and the presence of hydrops fetalis treated with steroid or surgery were included. Thirteen patients were treated with steroids, and 11 patients underwent open fetal surgery. In the steroid group 12 (92%) of 13 fetuses survived to delivery versus 9 (82%) of 11 in the open fetal surgery group. Only 5 (56%) of 9 of the patients in the open fetal surgery group survived to neonatal discharge compared to 10 (83%) of 12 in the steroid group.

Conclusions: In the present retrospective study, improved survival was seen in fetuses with hydrops fetalis and predominantly microcystic CPAM treated with steroids when compared with open fetal surgery. Steroids should be considered for first-line therapy in these cases.

Congenital cystic adenomatoid malformations (CCAM), now more correctly referred to as congenital pulmonary airway malformations (CPAMs) are rare lesions usually diagnosed during routine prenatal ultrasound. CPAMs are characterized by abnormal bronchopulmonary development [1]. The natural history of CPAMs are likely related to their growth pattern during embryologic development [2]. Lesion growth may increase or plateau during gestation. The size of the lesions and the variable growth behavior results in either the absence of symptoms or the development of non-immune fetal hydrops fetalis. Once fetal hydrops fetalis develops, mortality can reach 100% [3-5].

Treatment with steroids has been reported for microcystic CPAMs. Previous studies have shown a benefit with the use of steroids with survival to birth exceeding 85% [6-8]. Open fetal resection has also been used in cases of large CPAMs with hydrops fetalis, and this has been shown to improve outcome [9-11]. Although outcome is improved in these cases, open fetal surgery carries with it risks of intraoperative and postoperative complications. These complications include fetal bradycardia and demise, postoperative

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chorioamnionitis, amniotic fluid leakage, membrane separation and preterm labor [12].

We previously reported our experience with the use of steroids for microcystic CPAMs with hydrops fetalis [7]. Here we report our results of a retrospective comparison of steroids versus open fetal surgery in the treatment of predominantly microcystic CPAMs with hydrops fetalis.

1. Methods

This is an institutional review board–approved retrospective study of all patients referred to the University of California, San Francisco for the diagnosis of CPAM from 1997 to 2010. There was a total of 347 referrals for CPAM during this time. The fetuses from the surgery group were from the time period 1997 to 2001 and the fetuses from the steroid group were from 1997 to 2010. Fetuses with predominantly microcystic CPAM with the presence of hydrops fetalis who were treated with either steroids or surgery were included in the cohort. Microcystic CPAM was defined as >50% echogenic lesions on prenatal ultrasound. Hydrops fetalis was noted by the presence of 2 or more of the following: ascites, integumentary edema, pleural effusion, or placentomegaly. The fetuses that met these criteria were further subdivided into (1) open fetal resection or (2) maternal administration of a single course of betamethasone.

Data on patient characteristics included gestational age at diagnosis, CPAM volume to head circumference ratio (CVR) at diagnosis and upon resolution of hydrops, survival to birth, number of neonates on ventilator support at birth, resolution of hydrops, and time to resolution of hydrops. CVR was calculated by dividing the volume of the mass (length × width × height × 0.52) by the head circumference.

2. Results

A total of 24 pregnancies met the inclusion criteria for sonographically diagnosed predominantly microcystic CPAM with hydrops fetalis. Of the 24, 11 underwent open fetal resection of the CPAM and 13 received a single course of betamethasone. Before 2001, fetuses with microcystic CPAM and hydrops were offered surgery. The patients offered surgery were given steroids in preparation for fetal surgery. In this group, there were 3 patients that subsequently refused surgery. After 2001, all patients with microcystic CPAM and hydrops fetalis were offered steroids first.

In the steroid group 12 (92%) of 13 fetuses survived to delivery. Furthermore, 10 (82%) of 12 survived to neonatal discharge (Table 1). There were signs of hydrops in all of the patients in this cohort, and 10 (77%) of 13 had resolution of hydrops before delivery. The mean time to resolution of hydrops was 28 days. The single fetus that did not survive to delivery died at 27 weeks of gestation. The 2 patients who did not survive to neonatal discharge died on day of life 1. Both of these patients were delivered after spontaneous premature rupture of membranes at 24 weeks gestation. All of the non-survivors did not have resolution of hydrops.

In the open fetal surgery group, 9 (82%) of 11 fetuses survived to delivery and 5 (56%) of 9 survived to neonatal discharge (Table 1). In contrast to the steroid group, only 2 (18%) of 11 had resolution of hydrops. The mean time to resolution of hydrops was 26 days. The 2 fetuses that had resolution of hydrops survived to neonatal discharge. One of these patients did not require ventilator support. The 2 fetuses that did not survive until delivery died during attempted fetal resection. Most fetuses that did survive to delivery required ventilator support, 8/9 (89%). This is in contrast to only 1 (8%) of 12 in the steroid group. The remainder of neonates who did not survive to discharge died from respiratory failure.

There was no significant difference in CVR between the steroid versus the open fetal surgery group at the time of treatment. The mean CVR was 2.95 ± 0.31 for the surgery group and 2.68 ± 0.29 for the steroid group. In addition, no difference in CVR was seen in the nonsurvivors between the 2 groups. The mean decrease in CVR was 0.9 cm^2 ± 0.23 for the steroid group. The mean time between surgery and birth was lower than the time interval between steroid treatment to birth (35.7 ± 9.5 days vs. 87.6 ± 12.6 days, $P < .01$) (Fig. 1). The mean gestational age at delivery was 34 weeks (range, 24-40) for the steroid group and 31 weeks (range, 28-36) for the open fetal surgery group.

Table 1 Primary and secondary endpoints comparing steroid versus open fetal surgery in fetuses with predominantly microcystic CCAM

	Steroids	Open fetal surgery	*P*
Mean gestational age at delivery (wk)	34	31	<.05
% Survival to birth	12/13 (92%)	9/11 (82%)	.58
% Survival to neonatal discharge	10/12 (83%)	5/9 (56%)	.33
% Resolution of hydrops	10/13 (77%)	2/11 (18%)	<.05
CVR (cm^2) total	2.68 ± 0.29	2.95 ± 0.31	.55
CVR (cm^2) non-survivors	3.66 ± 0.66	3.02 ± 0.40	.42
Mean gestational age at treatment (wk)	23 (20-26)	24 (20-29)	.49
Ventilator requirement	1/12 (8%)	8/9 (89%)	<.001