Neonatal Outcomes of Prenatally Diagnosed Congenital Pulmonary Malformations

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**ABBREVIATIONS**

CPM—congenital pulmonary malformations

CVR—congenital pulmonary malformation volume ratio

IQR—interquartile range

ROC—receiver operating characteristic

**KEY WORDS**

congenital cystic adenomatoid malformation, lung development, sequestration

**WHAT’S KNOWN ON THIS SUBJECT:** Congenital pulmonary malformations are mostly identified prenatally. At birth, some children develop respiratory distress, which may be sufficiently severe to require mechanical ventilation and immediate surgery. The factors predictive of neonatal respiratory distress are not well defined.

**WHAT THIS STUDY ADDS:** Malformation volume and prenatal signs of intrathoracic compression are significant risk factors for respiratory complications at birth in fetuses with pulmonary malformations. In such situations, the delivery should take place in a tertiary care center.

**abstract**

**BACKGROUND AND OBJECTIVE:** Congenital pulmonary malformations (CPM) are mostly recognized on prenatal ultrasound scans. In a minority of cases, they may impair breathing at birth. The factors predictive of neonatal respiratory distress are not well defined, but an understanding of these factors is essential for decisions concerning the need for the delivery to take place in a tertiary care center. The aim of this study was to identify potential predictors of respiratory distress in neonates with CPM.

**METHODS:** We selected cases of prenatal diagnosis of hyperechoic and/or cystic lung lesions from RespiRare, the French prospective multicenter registry for liveborn children with rare respiratory diseases (2008–2013). Prenatal parameters were correlated with neonatal respiratory outcome.

**RESULTS:** Data were analyzed for 89 children, 22 (25%) of whom had abnormal breathing at birth. Severe respiratory distress, requiring oxygen supplementation or ventilatory support, was observed in 12 neonates (13%). Respiratory distress at birth was significantly associated with the following prenatal parameters: mediastinal shift (P = 0.003), polyhydramnios (P = 0.05), ascites (P = 0.005), maximum prenatal malformation area (P = 0.001), and maximum congenital pulmonary malformation volume ratio (CVR) (P = 0.001). Severe respiratory distress, requiring oxygen at birth, was best predicted by polyhydramnios, ascites, or a CVR > 0.84.

**CONCLUSIONS:** CVR > 0.84, polyhydramnios, and ascites increased the risk of respiratory complications at birth in fetuses with CPM, and especially of severe respiratory distress, requiring oxygen supplementation or more intensive intervention. In such situations, the delivery should take place in a tertiary care center. *Pediatrics* 2014;133: e1285–e1291

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(Continued on last page)
Congenital pulmonary malformations (CPM) are mostly identified prenatally, during routine second-trimester ultrasound examinations. They include various histologic entities, with congenital cystic adenomatoid malformations being the most frequent. The prevalence of this malformation in France, estimated by using European Surveillance of Congenital Anomalies network data, is between 1.34 and 1.94 per 10,000 births (2008–2011 data). The large volume of these malformations may lead to fetal pulmonary and mediastinal compression, with implications for fetal prognosis. At birth, most children with CPM are asymptomatic, but some develop respiratory distress that may be sufficiently severe to require mechanical ventilation and immediate surgery. Many of the previous studies evaluating neonatal respiratory morbidity in children with CPM used crude indicators, such as mortality or the need for immediate surgery. Only a few have focused on the prevalence of respiratory symptoms at birth, and the findings of these studies are often limited by an imprecise definition of the respiratory symptoms, the use of a single-center retrospective design, or a small number of patients. The heterogeneity of these studies probably contributed to the wide range of percentages of neonates with respiratory distress reported (13%–49%). Very few of these studies attempted to identify factors predictive of respiratory symptoms at birth, and conflicting results have been published concerning the predictive value of the size of the antenatal mass. Some of these studies have identified congenital pulmonary malformation volume ratio (CVR) as a significant predictor of neonatal respiratory distress, but discordant cutoff points have been proposed. Nevertheless, an understanding of these predictors is essential to optimize the care pathway after the prenatal diagnosis of lung malformations and to ensure that the appropriate level of care is provided at delivery. In 2008, a prospective registry of children born with CPM was established in France, under the auspices of the national health authority. A national Internet-linked based database (RespiRare) was established to record patient information and for the prospective collection of clinical data. This platform ensures the consistent input of numerous parameters and should provide reliable answers to unresolved issues. In the present study, we assessed the frequency of neonatal respiratory symptoms in children with CPM, with the aim of identifying the key prenatal parameters predictive of neonatal respiratory distress.

**METHODS**

**Study Design**

The RespiRare database for rare respiratory diseases in children was established in France to record extensive information on all forms of rare pediatric lung diseases, including CPM. A complete description of this database has been published elsewhere. Briefly, the reference center for these conditions and 8 affiliated university hospitals are located throughout France. Data for liveborn children are prospectively entered into the database via a secure Internet protocol and a Web interface. The database and data collection methods were approved by the French national data protection authorities (the National Commission on Data Processing and Liberties and the Advisory Committee on Information Processing concerning Health Research). Informed consent is obtained from parents before the inclusion of data for their children in the database. Data quality is ensured by the users entering the data and a database scientific committee that regularly reviews proposed cases for inclusion. A data manager and a technical team are responsible for quality control, the monitoring of data consistency and duplication, and the transfer of data, if required. We extracted, from this database, the files of patients born between January 2008 and March 2013 with a prenatal diagnosis of hyperechoic and/or cystic lung lesions. We identified 89 patients from 9 centers. This study was approved by the institutional review board of the French Respiratory Society (CEPR0 2013-016).

**Data Collection**

The prenatal and neonatal data available were: gestational age at diagnosis; appearance of the morphologic lesion, including its size, location, and type; associated abnormalities, such as mediastinal shift, polyhydramnios (defined as an amniotic fluid index greater than the 90th percentile for gestational age), or hydrops; gestational age at delivery and birth weight; and neonatal respiratory status, including tachypnea (defined as a respiratory rate >60 inspirations per minute), chest wall retraction, and need for oxygen supplementation or for ventilation. Lesions were classified as hyperechoic, cystic (diameter >5 mm), or both. Prenatal measurements of the size of the malformation are not recorded in the RespiRare database. At 1 center (Necker-Enfants Malades), which accounted for the largest number of patients (n = 53), data for malformation volume were extracted from the database of the Obstetrics Department, which was attended by the mothers during their pregnancies. Reliable data were obtained for 50 of the 53 patients. At this center, examinations were conducted with a Voluson 730 or VE8 ultrasound machine (GE Medical Systems, Zipf, Austria). The length, height, and width of the CPM were recorded, as previously described. The maximum area reached during pregnancy was calculated from the 2 largest dimensions. CPM volume was estimated with the formula for...
a prolate ellipse (length × height × width × 0.52). CVR was obtained by dividing the CPM volume by head circumference. Some pregnant women were not followed up at this tertiary center until the end of their pregnancy. Regression of the CPM at the end of the third trimester could be evaluated on ultrasound scans in 39 cases. An apparent regression >50% was considered significant.

**Statistical Analysis**

Quantitative variables are presented as median and interquartile range (IQR), corresponding to the difference between the third (75% of the distribution) and first (25% of the distribution) quartiles. Comparisons among groups were conducted by using the nonparametric Mann-Whitney test. Qualitative variables were expressed as percentages, and χ² tests were used for comparisons. In the primary analysis, we compared neonates with no respiratory distress with neonates displaying respiratory distress. Respiratory distress was defined as the presence of at least 1 respiratory symptom at birth: tachypnea (>60 inspirations per minute), chest wall retraction, oxygen supplementation, or a need for ventilation. In the secondary analysis, we considered the subgroup of neonates with more severe respiratory distress, defined as a need for oxygen supplementation or ventilation. This subgroup was compared with neonates not requiring oxygen at birth (ie, neonates with mild or no symptoms). P < .05 was considered to indicate a statistically significant difference. The performance of the CVR for predicting neonatal symptoms was investigated by plotting receiver operating characteristic (ROC) curves. Variables identified as significantly associated with respiratory distress were then entered into a multivariate logistic regression model. Because most of the cases originated from a single center (Necker-Enfants Malades), we evaluated a potential center effect by comparing the results obtained at this center with those obtained elsewhere.

**RESULTS**

**Prenatal Progression**

Eighty-nine fetuses with CPM were evaluated between February 2008 and April 2013. Median gestational age at diagnosis was 22.0 weeks. The malformation was unilateral in all cases and right-sided in 46 cases (52%). The lesions were cystic in 50 cases (56%), hyperechoic in 23 cases (26%), and mixed in 16 cases (18%). Doppler ultrasound scans identified a systemic vascular supply in 17 cases (19%). One or more signs of compression were detected on ultrasound scans in 33 cases (37%), including mediastinal shift (n = 32), polyhydramnios (n = 13), and/or ascites (n = 5). Hydrops was observed in 2 cases. The large volume of the malformation led to prenatal steroid treatment and/or thoracoamniotic drainage in 4 cases, including the 2 cases of hydrops. In the subgroup of cases for which CPM dimensions were available (n = 50), the lesion reached its maximal size at a median gestational age of 30.0 (IQR: 24.5–32.6) weeks. The median maximum surface area was 7.5 cm² (IQR: 3.8–13.8), and the median maximum volume was 11.8 cm³ (IQR: 4.5–23.4). Median CVR was 0.41 (IQR: 0.16–0.94). The CVR was <1.6 in all but 6 cases. The malformation size was reduced by more than one-half before birth in 18% of children.

**Neonatal Outcome**

Median gestational age at birth was 39.0 (IQR: 38.2–40.0) weeks (range: 30.0–42.0 weeks), and median birth weight was 3.4 (IQR: 2.9–3.6) kg (range: 1.9–4.5 kg). Sixty percent of neonates were male. Preterm birth <37 weeks occurred in 7 cases. Nine children had associated malformations (intratelial communication, n = 3; pectus excavatum, n = 2; uretero hydronephrosis, n = 4; hiatal hernia, n = 1). None of these associated malformations was considered sufficiently severe to contribute to respiratory symptoms at birth. Abnormal breathing at birth was observed in 22 cases (25%), characterized by tachypnea (n = 20) and/or chest wall retraction (n = 15) (Fig 1). Twelve of these children (13% of the total population) required oxygen supplementation, together with noninvasive ventilation in 4 cases and conventional mechanical ventilation in 6 cases. Early surgery (ie, before the end of the first month) was required in 5 children, on days 1 (n = 2), 3, 9, and 23. Two neonatal deaths occurred. One of the children who died had voluminous sequestration with prenatal compression but no hydrops. His maximum CVR was 3.2. Severe respiratory distress was observed at birth, requiring immediate intubation, and surgical removal on day 1. This child died on day 4. The second child had a voluminous cystic lesion with hydrops. Prenatal thoracoamniotic shunting was complicated by preterm birth at 30 weeks. This child died on day 1.

Several characteristics were significantly associated with neonatal respiratory distress (Table 1). Neonates with respiratory distress had a slightly lower gestational age at birth, but the difference did not reach significance. Neonates with respiratory distress were more likely to present with at least 1 prenatal sign of compression (P < .0001). Prenatal mediastinal shift, polyhydramnios, and ascites were associated with a significantly higher rate of neonatal respiratory distress (P = .0003, P = .05, and P = .0005, respectively). Consistent with this finding, prenatal maximum surface area, maximum volume, and maximum CVR were significantly higher in neonates with respiratory distress (P = .001 for all 3 parameters). When adjusted by using gestational age at delivery, the
association of any compression sign with neonatal respiratory distress remained highly significant (adjusted odds ratio: 6.9 [95% confidence interval: 2.2–20.9]; P < .001). This result was independent of recruiting center.

Limitation of the analysis to the subgroup of newborns with more severe respiratory distress requiring oxygen supplementation (n = 12) resulted in similar results (Table 2). The need for oxygen at birth was significantly associated with any prenatal sign of compression (P < .008), mediastinal shift (P < .04), polyhydramnios (P < .005), ascites (P = .0001), maximum prenatal malformation area (P < .003), maximum prenatal malformation volume (P < .005), and maximum CVR (P = .007). ROC curve analysis demonstrated that a CVR cutoff of 0.84 yielded the highest significance, with a sensitivity of 0.87 and a specificity of 0.81 (P = .0006). The area under the ROC curve was 0.804. Apparent regression of the CPM during the third trimester had no effect on oxygen requirement at birth. Polyhydramnios, ascites, and maximum CVR >0.84 were more reliable predictors of oxygen requirement at birth than mediastinal shift (Fig 2). For predicting the need for oxygen at birth, specificity was highest for the prenatal detection of polyhydramnios and/or ascites, whereas sensitivity was highest for a maximum CVR >0.84 (Table 3).

**TABLE 1** Prenatal Characteristics of Cases, Divided Into 2 Subgroups Based on the Occurrence of Respiratory Symptoms at Birth

<table>
<thead>
<tr>
<th>Characteristic</th>
<th>All Cases (N = 89)</th>
<th>Cases Without Neonatal Respiratory Distress (n = 67)</th>
<th>Cases With Neonatal Respiratory Distress (n = 22)</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Prenatal appearance on ultrasound scan</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Cystic</td>
<td>50 (56)</td>
<td>36 (54)</td>
<td>14 (64)</td>
</tr>
<tr>
<td>Hyperechoic</td>
<td>25 (28)</td>
<td>19 (28)</td>
<td>4 (18)</td>
</tr>
<tr>
<td>Mixed</td>
<td>16 (18)</td>
<td>12 (18)</td>
<td>4 (18)</td>
</tr>
<tr>
<td>Associated systemic vascularization*</td>
<td>17 (20)</td>
<td>14 (21)</td>
<td>3 (14)</td>
</tr>
<tr>
<td>Maximum surface area, cm²⁺</td>
<td>7.5 (3.8–13.8)</td>
<td>6.1 (3.4–10.1)</td>
<td>14.2 (8.4–20.1)*</td>
</tr>
<tr>
<td>Maximum volume, cm³⁺</td>
<td>11.8 (4.5–23.4)</td>
<td>8.1 (3.1–15.5)</td>
<td>27.4 (13.4–45.1)*</td>
</tr>
<tr>
<td>Maximum CVR, cm²⁺</td>
<td>0.41 (0.16–0.94)</td>
<td>0.26 (0.15–0.46)</td>
<td>1.01 (0.59–1.94)*</td>
</tr>
<tr>
<td>Apparent prenatal regression of CPM &gt;50%</td>
<td>7/39 (18)</td>
<td>3/27 (11)</td>
<td>4/12 (33)</td>
</tr>
<tr>
<td>Any sign of compression</td>
<td>35 (37)</td>
<td>17 (25)</td>
<td>16 (73)*</td>
</tr>
<tr>
<td>Mediastinal shift</td>
<td>32 (36)</td>
<td>17 (25)</td>
<td>15 (68)*</td>
</tr>
<tr>
<td>Polyhydramnios</td>
<td>13 (15)</td>
<td>7 (10)</td>
<td>6 (27)*</td>
</tr>
<tr>
<td>Ascites</td>
<td>5 (6)</td>
<td>0 (0)</td>
<td>5 (23)*</td>
</tr>
<tr>
<td>Hydrops</td>
<td>2 (2)</td>
<td>0 (0)</td>
<td>2 (9)</td>
</tr>
<tr>
<td>Gestational age at birth, wk</td>
<td>39.0 (38.2–40.0)</td>
<td>39.2 (38.3–40.0)</td>
<td>39.0 (38.2–38.7)</td>
</tr>
<tr>
<td>Birth weight, kg</td>
<td>3.4 (2.9–3.6)</td>
<td>3.4 (2.9–3.6)</td>
<td>3.4 (2.9–3.6)</td>
</tr>
<tr>
<td>Apgar score (1st minute)</td>
<td>10 (6–10)</td>
<td>10 (6–10)</td>
<td>8 (7.25–8)*</td>
</tr>
</tbody>
</table>

Data are presented as n (%), median (IQR), or n/N (%).

* One case for which data were unavailable in each subgroup.

* Data available for only 1 center (n = 35 without respiratory distress and n = 15 with respiratory distress).

* P < .05 between subgroups.
Both of these factors had high negative predictive values. Among children with no prenatal history of mediastinal compression, and with low CVR values, only 2 children required oxygen supplementation at birth, corresponding to a negative predictive value of 0.96 (Fig 3). However, these factors were not independent. A multivariate logistic regression analysis, including term, mediastinal shift, polyhydramnios or ascites, and CVR as independent variables, showed that only low CVR was significantly associated with absence of oxygen requirement at birth (odds ratio: 0.02 [95% confidence interval: 0.001–0.58]; \( P < .03 \)).

**DISCUSSION**

Our goal was to identify the predictors of neonatal respiratory distress in children with CPM. This study has several strengths with respect to other previously published studies on this topic. In particular, it was based on a prospective multicenter registry, allowing standardized data collection. As a result, we were able to analyze a large number of children and to avoid the biases associated with single-center retrospective studies. However, this study cannot be considered exhaustive, and it may not include all the cases occurring in France over the time period considered. This registry has been in use since 2008, but the participation of some centers is much more recent. Furthermore, only live births were considered. Despite these limitations, our results were found to be independent of the recruiting center, suggesting that the selection of the registered cases was unbiased. Furthermore, gestational age at diagnosis, gender distribution, and lesion type in our population were similar to those reported elsewhere.\(^{14-16}\)

A prenatal assessment of the risk of neonatal respiratory distress is important in mothers carrying fetuses with CPM, because decisions concerning the site at which the delivery takes place are dependent on this risk. Severe neonatal respiratory distress may occur, and deliveries should therefore occur at tertiary centers with a NICU and thoracic surgery expertise in the highest-risk cases. However, neonatal complications affect only a minority of children with CPM, and it is thus important to identify the principal prognostic factors. In our cohort, 25% of children had respiratory symptoms at birth, but only 13% required oxygen supplementation and 11% required ventilation support. Neonatal mortality was low in our cohort (2%). Previously published values are highly variable, probably due to major differences in the definitions used and case selection bias. Respiratory symptoms at birth were noted in 13% to 49% of cases in previous studies,\(^7-12\) with neonatal ventilatory support required.

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**TABLE 2** Prenatal Characteristics of Cases, Divided Into 2 Subgroups Based on the Requirement for Oxygen Supplementation at Birth

<table>
<thead>
<tr>
<th>Characteristic</th>
<th>Cases Without Oxygen Requirement at Birth ((n = 77))</th>
<th>Cases With Oxygen Requirement at Birth ((n = 12))</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cystic</td>
<td>40 (52)</td>
<td>109 (83)</td>
</tr>
<tr>
<td>Hyperechoic</td>
<td>22 (29)</td>
<td>1 (8)</td>
</tr>
<tr>
<td>Associated systemic vascularization(^a)</td>
<td>16 (21)</td>
<td>1 (8)</td>
</tr>
<tr>
<td>Maximum surface area, cm(^2)</td>
<td>6.8 (5.6–10.7)</td>
<td>16.0 (14.0–23.1)*</td>
</tr>
<tr>
<td>Maximum volume, cm(^3)</td>
<td>9.0 (4.1–17.1)</td>
<td>28.8 (25.7–51.7)*</td>
</tr>
<tr>
<td>Maximum CVR, cm(^2)</td>
<td>0.34 (0.16–0.64)</td>
<td>1.17 (0.96–2.07)*</td>
</tr>
<tr>
<td>Any sign of compression</td>
<td>24 (31)</td>
<td>9 (75)*</td>
</tr>
<tr>
<td>Mediastinal shift</td>
<td>24 (31)</td>
<td>8 (67)*</td>
</tr>
<tr>
<td>Polyhydramnios</td>
<td>8 (10)</td>
<td>5 (42)*</td>
</tr>
<tr>
<td>Ascites</td>
<td>1 (1)</td>
<td>4 (33)*</td>
</tr>
<tr>
<td>Hydrops</td>
<td>0</td>
<td>2 (17)*</td>
</tr>
<tr>
<td>Gestational age at birth, wk</td>
<td>39.2 (38.4–40.0)</td>
<td>38.8 (38.1–38.8)</td>
</tr>
<tr>
<td>Birth weight, kg</td>
<td>3.4 (3.0–3.6)</td>
<td>3.1 (2.8–3.6)</td>
</tr>
<tr>
<td>Apgar score (1st minute)</td>
<td>10 (8–16)</td>
<td>8 (6.75–8.25)*</td>
</tr>
</tbody>
</table>

* Data are presented as \(n\) (%), median (IQR), or \(n/N\) (%).
* Two cases for which data were unavailable in the subgroup without oxygen supplementation.
* Data available for only 1 center (\(n = 50\)).
* \(P < .05\) between subgroups.
in up to 22% to 25% of neonates. The incidence of severe symptoms at birth therefore seems to be lower in our series than in previous studies, probably because we studied an unselected population, with multicenter recruitment. It is also possible that improvements in the performance of antenatal ultrasound have made it possible to identify small malformations that previously went unnoticed, increasing the proportion of asymptomatic lesions at birth. It is not possible to evaluate the precise number of pregnancies with CPM that do not result in live births from our registry, but this rate can be estimated at ∼8% in France based on European Surveillance of Congenital Anomalies network data. This frequency is consistent with previously published evaluations, which ranged from 3% to 23%. Of interest, early surgical removal of the malformation (ie, within the first 30 days after birth) was required in only a minority of children with severe respiratory symptoms at birth. Spontaneous improvement was observed in 2 of the 6 children on conventional mechanical ventilation and in 3 of the 4 children with noninvasive ventilation. In these neonates, no other condition could have contributed to the respiratory distress. It is possible that adaptation to extraterine life in these children was transiently affected by delayed amniotic fluid clearance or some degree of pulmonary hypoplasia, both induced by the CPM.

The maximum prenatal size of the mass and its compressive effects were clearly related to neonatal outcome. Mediastinal shift was observed in a large number of fetuses, the rate of 36% obtained for our population being slightly lower than published rates ranging from 45% to 79%. Mediastinal shift has previously been identified as significantly associated with the need for postnatal surgery. In our series, prenatal mediastinal shift was significantly associated with respiratory distress at birth but was not predictive of the severity of symptoms. In neonates with respiratory distress, the prenatal detection of a mediastinal shift did not discriminate between infants requiring oxygen supplementation or ventilatory support and those not requiring such assistance. By contrast, the prenatal detection of polyhydramnios and/or ascites was found to be more specific for the prediction of severe respiratory symptoms at birth. These factors also had a high negative predictive value for the need for oxygen at birth. Polyhydramnios and ascites probably reflect larger malformations, with more severe mediastinal compression. This outcome was confirmed in children for whom malformation measurements were available. Maximum surface area and maximum CVR were significantly associated with neonatal respiratory distress and with oxygen requirement. Maximum CVR was the most sensitive risk factor for oxygen requirement at birth. It also had a high negative predictive value. The CVR was initially validated for predicting the risk of hydrops, but 2 previous studies, by Yong et al and by Ehrenberg-Buchner et al, suggested that it might also be of predictive value for postnatal outcome. Different cutoff points for maximum CVR were identified in these 2 studies: 0.56 and 1.0, respectively. The composite nature of the postnatal score used in the

<table>
<thead>
<tr>
<th>Prenatal Sign</th>
<th>Neonatal Symptom</th>
<th>Sensitivity</th>
<th>Specificity</th>
<th>PPV</th>
<th>NPV</th>
</tr>
</thead>
<tbody>
<tr>
<td>Polyhydramnios and/or ascites</td>
<td>Any symptom</td>
<td>0.41</td>
<td>0.90</td>
<td>0.56</td>
<td>0.82</td>
</tr>
<tr>
<td></td>
<td>Oxygen requirement</td>
<td>0.58</td>
<td>0.88</td>
<td>0.44</td>
<td>0.93</td>
</tr>
<tr>
<td>Mediastinal shift</td>
<td>Any symptom</td>
<td>0.68</td>
<td>0.75</td>
<td>0.47</td>
<td>0.88</td>
</tr>
<tr>
<td></td>
<td>Oxygen requirement</td>
<td>0.67</td>
<td>0.80</td>
<td>0.25</td>
<td>0.93</td>
</tr>
<tr>
<td>Maximum CVR &gt;0.84</td>
<td>Any symptom</td>
<td>0.67</td>
<td>0.86</td>
<td>0.67</td>
<td>0.86</td>
</tr>
<tr>
<td></td>
<td>Oxygen requirement</td>
<td>0.87</td>
<td>0.81</td>
<td>0.47</td>
<td>0.97</td>
</tr>
</tbody>
</table>

NPV, negative predictive value; PPV, positive predictive value.
study by Yong et al, with the inclusion of complications occurring several months after birth, renders comparison with our results difficult. The study published by Ehrenberg-Buchner et al is more similar to ours, because it was limited to neonatal respiratory morbidity. The slightly lower CVR cutoff point that we identified may reflect differences in the proportions of children with severe respiratory symptoms. We found that higher values of CVR were associated with more severe respiratory distress, requiring at least oxygen supplementation, and the proportion of children with severe symptoms was not reported in the study by Ehrenberg-Buchner et al. Because these severe respiratory complications occur in a small minority of neonates with CPM, larger prospective studies will be required to define the optimal CVR cutoff point.

CONCLUSIONS

Maximum CVR and prenatal signs of intrathoracic compression are significant risk factors for respiratory complications at birth in fetuses with CPM. A CVR > 0.84, polyhydramnios, and ascites were significantly associated with more severe respiratory distress, requiring at least oxygen supplementation. In these situations, delivery at a tertiary care center should be planned.

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