reported [12]. Other prognostic factors have been described: mediastinal shift and polyhydramnios which, however, seem to be poor indicators of prognosis; the size of the mass, especially when occupying > 50% of the thorax, and pulmonary hypoplasia, which is very difficult to evaluate ultrasonographically [4]. The microcystic form of CAML is a predictor of the worst prognosis (but there is no general agreement). Although limited, our experience seems to confirm that CAML without polyhydramnios and/or hydrops has an excellent outcome. Moreover, spontaneous regression has also been recently reported in fetuses with polyhydramnios or ascites [12].

Laryngeal obstruction has been observed to produce a prenatal ultrasonographic and histological appearance similar to CAML [2, 6]. Therefore, it has been suggested that this association could explain the apparent resolution of some cases of CAML [12] and Dumez et al. [4] reported such prenatal unpredictable "disappearance" in approximately 21% of cases (3 of 14 cases). In our experience all those lesions decreasing in size during fetal life were microcystic. This lends support to the hypothesis of a possible correlation between the prenatal image and the clinical disappearance or resolution of the lesion. Maybe the microcystic appearance could be a "false image" related to a temporary laryngeal obstruction [2, 13] or a simple inhalation of vernix caseosa. In one case of our series, who underwent prenatal resolution, a remarkable amount of vernix caseosa was clearly evident in the amniotic fluid.

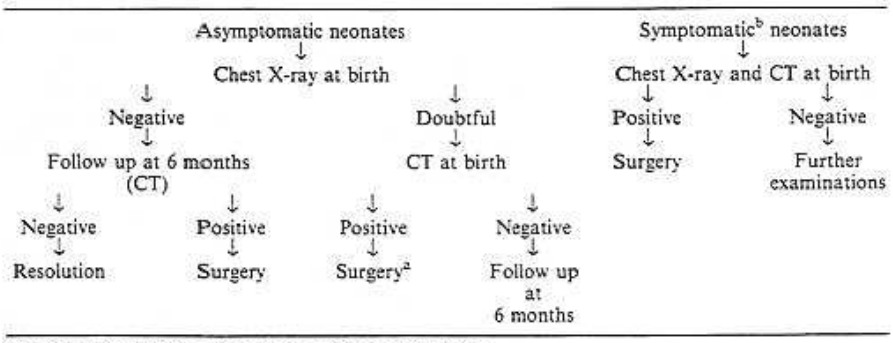
Prenatal therapy would be indicated in the presence of severe hydrops or a dramatically increasing mediastinal shift. In the case of a large cyst, drainage may be performed; however, fluid re-accumulation is not prevented. Thoraco-amniotic shunting, although not completely free of complications, has been associated with good outcome [3]. The role of fetal surgery by hysterotomy, especially considered for solid lesions, still remains to be evaluated. No fetuses in our series needed prenatal therapy.

A wide spectrum of postnatal clinical presentation of CAML has also been described ranging from acute respiratory distress or even neonatal death to mild respiratory symptoms during childhood. The postnatal diagnostic assessment includes Cxr and CT ± angiography. Surgical treatment requiring thoracotomy and lobectomy is frequently unavoidable, as in the case of four neonates in our series with positive Cxr, CT and clinical symptoms.

In the newborn, surgery is often performed as soon as the diagnosis is confirmed, even in asymptomatic patients [9]. This approach could be justified by the possible occurrence of a malignant change [8]; however, this "is analogous to prophylactic nephrectomy of multicystic renal disease" [12]. Therefore, in our opinion, the surgical approach is not justified at birth if the newborn is asymptomatic with either negative or positive Cxr. In fact, when there is no definitive evidence of a malignant change of the lesion, a postnatal disappearance has been reported [12]. Moreover, the prenatal image could be produced by a temporary laryngeal obstruction; thus, both the prenatal ultrasound and the neonatal radiological examination are not probative for CAML. The histological evaluation could provide evidence of the lesion. However, neither is it a routine procedure nor can it be useful in making a decision for surgery. In fact, it must be taken into account that a temporary laryngeal obstruction may produce a similar histological appearance [2, 6] and that CAML might resolve postnatally. Approximately 26% of the surviving infants in a series with prenatally diagnosed CAML did not undergo surgery because of either absence of symptoms or resolution of pathology [12]. Our experience (Table 2) suggests that surgery has to be performed at birth in the symptomatic (respiratory distress, polypnoea, altered blood gas analysis parameters, etc.) patients, but could be safely delayed (6 months) in the asymptomatic ones (Table 3).

For the same reasons, a CT at birth would be necessary only when preliminary to surgery. On the contrary, in asymptomatic neonates it is necessary at birth only when the Cxr is positive or suspicious for a pulmonary lesion. If the Cxr is negative, CT should be performed at 6 months of age for a better definitive assessment of the patient (Table 3).

Table 3 Algorithm proposed for postnatal management of neonates with prenatally diagnosed CAML



[a] Surgery may be delayed until 6 months in these cases
[b] Respiratory distress, polypnoea, reduced PaO_2, increased $PaCO_2$, etc

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