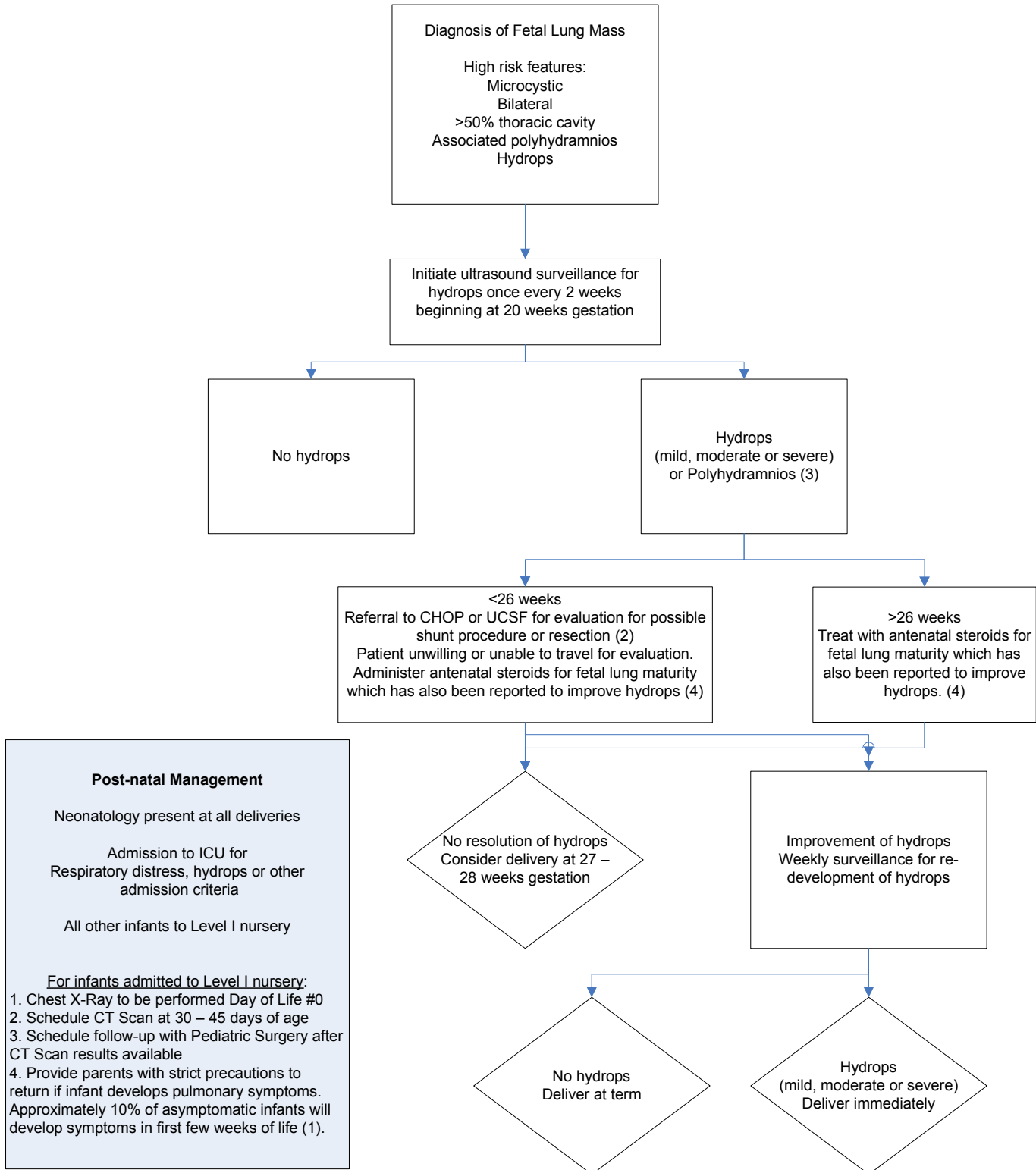


Congenital Cystic Adenomatoid Malformation and Pulmonary Sequestration Prenatal Management Algorithm



1. Sauvat F, Michel J, Benachi A, Edmond A, Revillon Y. Management of Asymptomatic Neonatal Cystic Adenomatoid Malformations. *J Pediatr Surg* 38:548-552, 2003.

29 cases of antenatally diagnosed CCAM in which infants were asymptomatic at birth were identified. Chest x-Ray performed within first days of life, follow-up CT scan performed around 45 days of life. 12 cases (41.3%) had normal CXR at birth, 3 (10.3%) of which subsequently developed clinical manifestations. CT scan was normal in 4 (13.8%) cases, only one of which had a normal CXR as well. CCAM vanished in 6 cases (1 antenatally, 3 at birth, 1 first months of life, 1 first years of life). 17 (58.6%) required surgical resection (3 for symptoms during neonatal period, 15 for asymptomatic infants with either large lesions >3 cm or liquid-filled CCAM predisposing to infection). Long term outcome is excellent. One child with asthma after surgical resection, all others asymptomatic.

2. Dommergues M, Louis-Sylvestre C, Mandelbrot L, Aubrey MC, Revillon Y, Jarreau PH, Dumez Y. Congenital adenomatoid malformation of the lung: When is active fetal therapy indicated? *Am J Ob Gynecol* 177: 953-8, 1997.

Case series of 33 patients. Ultrasound evaluation every 2 weeks. Aggressive management with invasive fetal therapy only considered in cases with hydrops or severe polyhydramnios (12/33). They offered percutaneous thoracoamniotic drainage with double-pigtail catheter only. Open fetal surgery was not offered. Two patients with severe poly/hydrops terminated their pregnancy. Thoracoamniotic shunting performed in 9 patients, 4 of whom survived. One patient with Type III/microcystic lesion and hydrops/poly was not a candidate for shunting, was managed conservatively and survived. 17 cases without acute polyhydramnios or hydrops were managed conservatively and survived.

3. Mahle WT, Rychik J, Tian ZY, Cohen MS, Howell LJ, Crombleholme TM, Flake AM, Adzick NS. Echocardiographic evaluation of the fetus with congenital cystic adenomatoid malformation. *Ultrasound Obstet Gynecol.* 16:620-4, 2000.

Echocardiogram performed on 41 consecutive fetuses with CCAM. Hydrops present in 15/41 (36.5%). Fetuses with hydrops demonstrated an increase in early ventricular filling, higher ratios of early ventricular filling to atrial contraction (E/A ratio) at mitral and tricuspid valves, and higher degree of reversal of flow in IVC with atrial contraction. Authors propose elevated CVP resulting from mass effect on great vessels as mechanism of hydrops. Authors suggest that echocardiographic criteria may become useful in the prediction of hydrops in fetuses with CCAM.

4. Tsao KJ, Hawgood S, Vu L, Hirose S, Sydorak R, Albanese CT, Farmer DL, Harriston MR, Lee H. Resolution of hydrops fetalis in congenital cystic adenomatoid malformation after prenatal steroid therapy. *J Pediatr Surg.* 38: 548-52, 2003.

Small case series of 3 fetuses with CCAM and mild to moderate hydrops at 24, 26 1/7, and 21 3/7 weeks gestation. All fetuses were given two doses of Betamethazone 12 mg IM 24 hours apart.

All three pregnancies demonstrated gradual resolution of hydrops with delivery at 42 6/7, 39 and 39 weeks gestation and all three were alive and well at the time of publication. Although mechanisms are speculative, the authors propose that steroid therapy may provide a boost to lung development which accelerates resolution of the lesion.