Trust Guideline for the Management of Congenital Thoracic Malformations

A clinical guideline recommended for use

For Use in:	Obstetric Department, Neonatal Intensive Care Unit (NICU), Buxton Ward, Radiology Department, Anaesthetic Department, Theatre staff
By:	Staff in the above areas
For:	The management of Congenital Thoracic Malformations both antenatally and postnatally
Division responsible for document:	Women and Children's Division
Key words:	Cystic adenomatoid malformation, Pulmonary sequestration, Antenatal, Ultrasound
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Assessed and approved by the:	Clinical Guidelines Assessment Panel (CGAP) If approved by committee or Governance Lead Chair's Action; tick here ☐
Date of approval:	19/08/2021
Ratified by or reported as approved to (if applicable)**:**	Clinical Safety and Effectiveness Sub-board
To be reviewed before: This document remains current after this date but will be under review	19/08/2024
To be reviewed by:	Authors
Reference and / or Trust Docs ID No:	1353
Version No:	3.2
Compliance links: *(is there any NICE related to guidance)*	None
If Yes - does the strategy/policy deviate from the recommendations of NICE? If so why?	N/A

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Version and Document Control:

Version Number	Date of Update	Change Description	Author
3.2	19/08/2021	Document reviewed, no clinical changes required.	Dr Chris Upton Mrs Ngozi Joy Nwokomo

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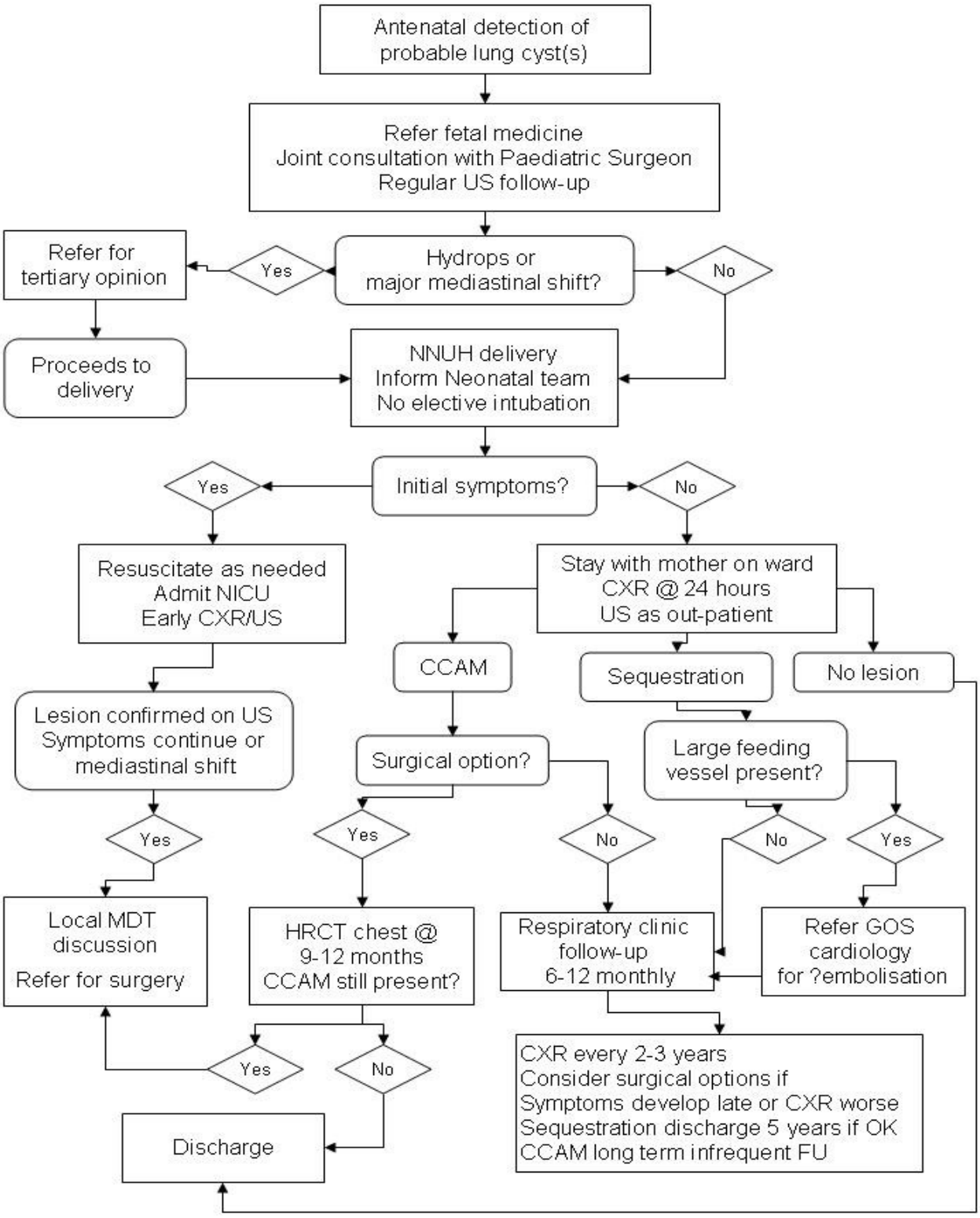
Quick reference guidelines



Brief reference guidelines – see algorithm above

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Objectives

To provide guidance for the optimal management of Congenital Thoracic Malformations.

Rationale

The guideline was written to try to produce some conformity in the investigation, management and follow up of Congenital Thoracic Malformations. The guideline has a strong evidence base but the literature available is strongly biased towards surgical intervention, even in asymptomatic children, and we try to present a more balanced view of the available evidence.

Broad recommendations

1. Antenatal management

Following the detection of suspected Congenital Thoracic Malformation (CTM) on routine antenatal ultrasound, the patient should be referred to Fetal Medicine for further evaluation. The main differential diagnosis is a diaphragmatic hernia. Colour-Doppler ultrasound should be performed to demonstrate any anomalous blood flow from the aorta to the lung malformation. The mother should be reviewed and counselled appropriately in a combined Obstetric/Surgical consultation including information on the possible diagnosis, likely prognosis and available monitoring and treatment options including the choice of fetal karyotype and need to deliver the child in Norwich rather than a neighbouring DGH.

Repeat ultrasound scans should be performed two to four-weekly to monitor the size and growth of the CTM as well as the presence of any mass effects such as hydrops fetalis, ascites, pleural or pericardial effusion and mediastinal shift [1]. If there is evidence of mass effect monitoring ultrasound can be performed more frequently. Fetal MRI may help to distinguish CTM from diaphragmatic hernia if there is doubt, but it is an expensive tool and requires referral to Sheffield.

During antenatal monitoring, the progression of mass effects may be an indication for referral for a tertiary opinion regarding fetal intervention, although there is little evidence for the efficacy of interventions such as thoraco-amniotic shunting. There is no advantage in being delivered early or via Caesarean section unless there are obstetric indications. The neonatal intensive care team should be notified during pregnancy via the Paediatric Alert System and again as soon as possible when the mother is admitted for delivery. Specific information about complications such as hydrops and mediastinal shift or if the lesion is small and regressing should be provided.

2. Postnatal management

A Paediatric ST or ANNP should be present at delivery in case of early respiratory distress and need for resuscitation, although many babies are asymptomatic at birth and do not need separating from their parents nor admission to NICU. This is because many CTMs regress in size towards term. Criteria for active resuscitation should be exactly the same as for any newborn baby.

Whereas in diaphragmatic hernia elective intubation is necessary, in CTM this is not the case and elective intubation should be avoided. It may in fact cause paradoxical deterioration by distending a lung cyst further and provoking mediastinal shift. Babies that require resuscitation should be admitted to NICU and a chest X-ray performed soon to assess the degree of any cystic lesion present. Asymptomatic babies should be examined but if the examination is normal they should be left with parents with appropriate observations on the post-natal ward and a chest X-ray performed at 24 hours of age. The X-ray is easier to interpret at this age after the clearance of lung fluid. Plain chest X-rays tend to underestimate the extent of any underlying CTM and all infants should be evaluated further after discussion with Dr Pickworth or Dr MacIver in radiology and an ultrasound is usually preferred. Ultrasound in asymptomatic babies can be arranged as an outpatient a few weeks after discharge. If there is no evidence of any residual in congenital cystic adenomatoid malformation (CCAM) or sequestration on ultrasound then the infant can be safely discharged from follow up.

a) Symptomatic infants or those with mediastinal shift on X-ray

Infants with early symptoms of respiratory distress or with mediastinal shift on X-ray are likely to require early resection of their lesion. The next radiological investigation of choice is usually an Ultrasound scan of the chest – to discuss with a radiologist. The aim of the ultrasound is to distinguish (CCAM), which is an isolated lung lesion, from pulmonary sequestration, where the lesion has an arterial blood supply from the systemic not pulmonary circulation. Colour-Doppler ultrasound is the initial investigation of choice for distinguishing the two. Sequestration can cause symptoms via high output cardiac failure from the aberrant arterial supply as well as by mass effect.

Babies in this group are likely to require surgical correction of their lesion and should be discussed with a paediatric surgeon with a thoracic interest (Mr Minocha or Mr Kulkarni). The surgery is often performed in a paediatric cardio-thoracic centre such as in Great Ormond Street (GOS) although may be operated on locally if appropriate surgical, anaesthetic and neonatal support is available. The urgency of such intervention will depend on the severity of the respiratory distress and is clearly most urgent if the infant is ventilator dependent as a consequence of the CTM. Discuss with the centre whether they prefer a high resolution CT scan of the chest to be performed in Norwich and cabled across to them on PACS, or whether they would prefer their own CT on arrival.

Infants that have had surgical correction of their CTM should be referred to Dr Upton /Dr Kavanagh for long term follow up in the Childrens' Respiratory Clinic, initially in six months with frequency of appointments reducing with time. There is an extremely small risk of malignant change in CCAM but there is no evidence that surgery removes this risk, so long term but infrequent follow up with occasional repeat chest X-rays is sensible.

b) Asymptomatic infants, no mediastinal shift

i. CCAM detected on initial ultrasound

This is the group in which there is the most controversy in terms of management. Many groups advocate surgery for all but there is good

evidence that CCAM detected on HRCT can resolve spontaneously in the first year of life [2], so early surgery before a year old in asymptomatic infants should not be recommended. In addition the quoted frequency of symptomatic CCAM of 1 in 25,000 births is vastly lower than the frequency of lesions detected by antenatal ultrasound in this paper, suggesting many lesions remain asymptomatic for many years.

Lesions may become infected and if they do then surgery may prove more difficult. However most of the published evidence relates to CCAM picked up clinically via infection and one cannot extrapolate that CCAM being followed up via antenatal detection that is then infected will produce the same difficulties. Indeed our experience locally is that those who develop recurrent infections after initial conservative treatment are very quick to consult and be referred for surgical intervention if required.

There are now approximately 50 international case reports over many years of malignant change developing in CCAMs, usually pleuropulmonary blastomas or carcinomas. These are mostly in adults but the youngest in a child of 13 months. However the presentation is usually with bilateral disease and/or pneumothorax or with a positive family history for blastomas. Most are not typical unilateral asymptomatic lesions. Additionally there have also been case reports of malignant change developing after resection of CCAM, so parents cannot be reassured that resection removes the risk or need for follow up.

The parents of infants with asymptomatic CCAMs should be counselled about the risks of both infection and malignancy and be able to make an informed decision about whether to be referred for surgery. We do not believe that early HRCT of the chest is required in asymptomatic patients whose parents opt for conservative management. Nor is early CT, for those who want surgery, sensible in view of the tendency to regress – they should be reassessed at 9 months to a year of age and if surgery the preference a CT arranged then to check for persistence of the lesion.

Whether parents opt for surgery or not this group should also be offered follow up in the Childrens' Respiratory Clinic as above.

ii. Sequestration detected on initial ultrasound

Sequestration may be intra or extra-pulmonary but it is rare for either form to connect to the airway and so respiratory infection is rare. Nor has malignancy been reported. Additionally 75% of sequestrations resolve with time so the majority can be managed safely conservatively. The main risk with sequestration is that of high output cardiac failure and the infant should be assessed carefully clinically for this. If the radiologist is concerned about the flow in the feeding vessel, then a full assessment should be made via referral to Dr Graham Derrick, Cardiologist from GOS, at his Norwich clinic. If high output cardiac failure is confirmed due to a large feeding vessel, many can be treated via embolisation at GOS using interventional cardiology techniques, rather than requiring open surgery. Scimitar syndrome is a variant of sequestration in the right lung resulting in pulmonary hypoplasia, infra-

diaphragmatic venous drainage to the vena cava from the sequestered lobe and surrounding normal lung. This can be sometimes seen or picked up from a CXR due to its characteristic appearance.

Initial follow up of sequestration, whether they have required intervention or not, is sensible to check for lack of progression or symptoms, either in cardiology or respiratory clinic. However, these children do not require long term follow up after the age of five years.

iii. Congenital hyperlucent lobe

This condition, previously known as congenital lobar emphysema, is not strictly a CTM but may be confused for one. It presents less commonly antenatally than true CTM and more likely as respiratory distress in the first few months of life. There is hyperlucency of the affected lobe, most commonly an upper lobe, which may compress adjacent lung. Those with severe symptoms will require surgical intervention with lobectomy, but there is increasing evidence that resolution may occur in those with milder symptoms with conservative management [3].

Clinical audit standards

- Appropriateness of initial radiology, i.e. chest X-ray at appropriate time, ultrasound or CT only when indicated.
- All parents to be counselled clearly regarding the risks of infection and malignancy.
- Symptomatic babies to have appropriate surgery and follow up arranged.
- Parents of asymptomatic infants to be offered a choice of surgical or conservative approach with appropriate follow up either way.

Summary of development and consultation process undertaken before registration and dissemination

The authors listed above drafted the guidelines. It has been circulated to the Paediatric Thoracic Group, Departments of Paediatric Surgery and Paediatric Medicine and Neonatology (consultants, junior medical staff, nursing staff on Buxton Ward and CAU), consultant thoracic surgeons, consultants in fetal medicine, consultant paediatric anaesthetists and paediatric radiologists for comments.

This version has been endorsed by the Clinical Guidelines Assessment Panel.

The guideline was reviewed in August 2021 and no clinical changes were necessary.

Distribution list/ dissemination method

Trust intranet, NICU, CAU, Buxton Ward and Antenatal clinic

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References/ source documents

1. Pumberg W, Hörmann M, Deutinger J, Bernaschek G, Bistricky E, Horchner E. Longitudinal observation of antenatally detected congenital lung malformations (CLM): natural history, clinical outcome and long-term follow up. Eur. J Cardiothoracic Surg 2003; **24**: 703-711
2. Calvert JK, Boyd PA, Chamberlain PC, Syed S, Lakhoo K. Outcome of antenatally suspected congenital cystic adenomatoid malformation of the lung: 10 years' experience 1991-2001. Arch Dis Child Fetal Neonatal Ed 2006; **91**:F26-F28
3. Kovacevic A, Schmidt KG, Nicolai T, Wisbauer M, Schuster A. Two further cases supporting nonsurgical management in congenital lobar emphysema. Klin Padiatr 221; **4** 2009: 232-6.