Surgical versus Conservative Management for Asymptomatic Congenital Cystic Lung Malformations in Children

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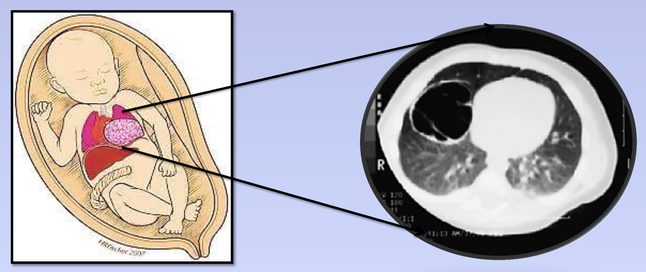
Background

- Congenital cystic adenomatoid malformations (CCAM) is relatively rare congenital anomaly where a portion of the lung is replaced by a piece of non-functioning cystic lung tissue.
- The presentation of CCAM ranges from respiratory distress at birth to entirely asymptomatic lesions [1, 2].
- Surgical intervention has long been recommended for management of symptomatic lesions due to the associated morbidity and mortality[3].
- When cases of known cystic lung disease are asymptomatic the ideal management is less defined.
- An increasing proportion of neonates are presenting asymptomatically due to identification on antenatal ultrasound, presenting a significant challenge to pediatric surgeons.



Rationale and Objective

- Asymptomatic CCAM is an increasingly prevalent challenge in pediatric general surgery
- Debate persists over the optimal management of asymptomatic lesions
- Prior reviews of CCAM management exist but are largely non-systematic in nature
- To the best of our knowledge, no high quality systematic reviews comparing management approaches for CCAM currently exist.

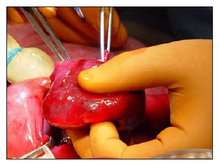
Our objective was to perform a systematic review and meta-analysis to evaluate and compare the risks of elective surgery versus expectant management for cases of asymptomatic CCAM in children

Arguments in favour of surgical management

- Delaying surgery until symptoms develop may result in a more complicated procedure with worse surgical outcomes
- Resolution on imaging, supporting conservative management, may be attributable to growth of healthy lung tissue around the lesion and not true regression
- Whether the prevalence of cases of resolution is great enough to justify observation given the morbidity associated with the condition is undetermined

Arguments in favour of surgical management

- Rates of symptom development are reported as low as 3% or as high as 85% therefore it is controversial whether it is worth subjecting patients to potentially unnecessary surgical risk
- Resolution of CCAM lesions is estimated to occur in 4-13% of cases



Methods

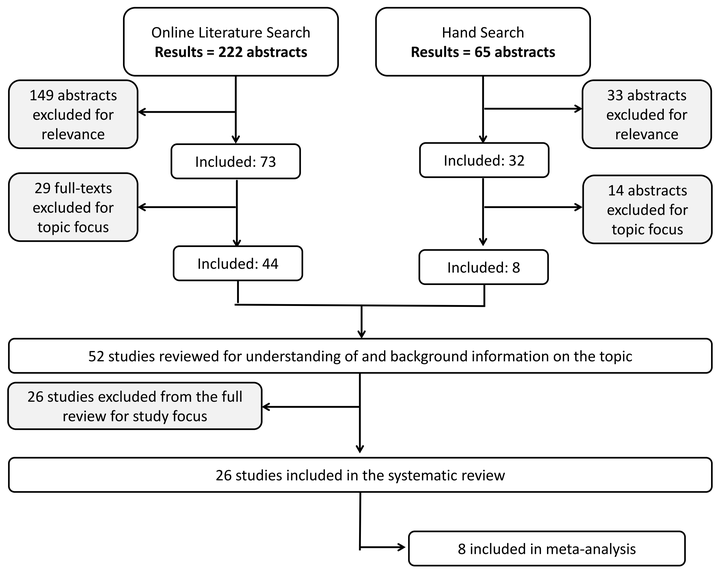
Search
- Electronic search of the Cochrane Central Register of Controlled Trials (CENTRAL), MEDLINE, EMBASE), CINAHL and PUBMED to identify relevant primary studies and reviews on the treatment of CCAM in neonates and children.

Hand Search
- The search was expanded by performing a hand search of the references of all included studies.

Review
- Both abstracts and full texts of retrieved articles were reviewed for inclusion in a two-tier review process by two independent reviewers . Where consensus on an article could not be reached a third reviewer was consulted

Study exclusion criteria:
- Studies published in a language other than English
- Editorials, case studies and letters
- Studies that did not describe management, described antenatal management only or surgical management only
- Studies that had a primarily adult study populations.

Figure 1. Study Flow Diagram



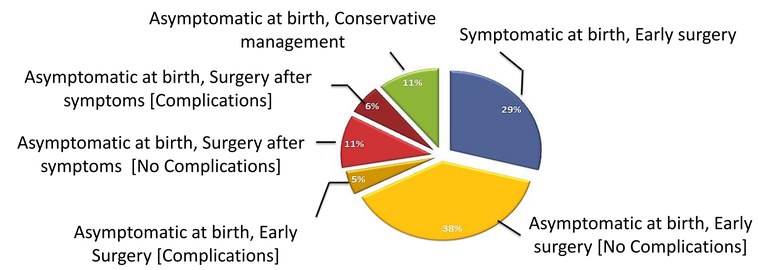
Quality Assessment

- Two researchers independently assessed each study using the Methodological Index for Non-Randomized Studies (MINORS)
- All studies but one were retrospective in nature
- No studies employed a blinding technique for data collection
- No studies did a prospective calculation of study size
- Methodological biases are reflected in total MINORS scores
  - Range: 7 to 16
  - Median: 8.5
- - Maximum score for non-comparative studies is 16, maximum score for comparative studies is 24

Results

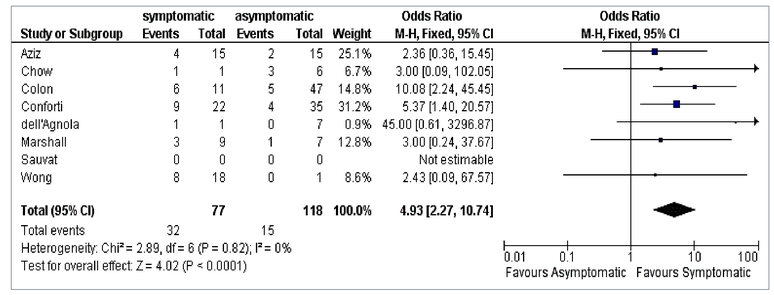
- 262 patients were symptomatic at birth requiring immediate surgery
- 641 patients were asymptomatic at birth and were managed surgically before or after symptom development or were managed conservatively (1 patient was lost to follow up before treatment)
- 389 of the asymptomatic patients underwent early elective surgery with 43 experiencing post-operative complications
- 151 of the asymptomatic patients did not undergo early surgery and went on to develop symptoms at a mean 14.6 months. These patients underwent surgery after developing symptoms with 54 experiencing post-operative complications
- 99 patients were managed conservatively and all remained symptom free at a mean follow up of 37 months.

Figure 2. Percentage of study population who received each management course



- Meta-analysis was performed on 8 of the included studies. Our pooled estimate from the analysis showed that total morbidity (as defined by number of patients who experienced post-operative complications) was higher when surgery was performed after symptom development (OR: 4.93 [2.27, 10.74], p<0.0001).

Figure 3. Meta Analysis Results



Study or Subgroup	symptomatic Events	Total	asymptomatic Events	Total	Weight	Odds Ratio M-H, Fixed, 95% CI
Aziz	4	15	2	15	25.1%	2.36 [0.36, 15.45]
Chow	1	1	3	6	6.7%	3.00 [0.09, 102.05]
Colon	6	11	5	47	14.8%	10.08 [2.24, 45.45]
Conforti	9	22	4	35	31.2%	5.37 [1.40, 20.57]
dell'Agnola	1	1	0	7	0.9%	45.00 [0.61, 3296.87]
Marshall	3	9	1	7	12.8%	3.00 [0.24, 37.67]
Sauvat	0	0	0	0		Not estimable
Wong	8	18	0	1	8.6%	2.43 [0.09, 67.57]
Total (95% CI)		**77**		**118**	**100.0%**	**4.93 [2.27, 10.74]**
Total events	32		15			

Heterogeneity: Chi² = 2.89, df = 6 (P = 0.82); I² = 0%
Test for overall effect: Z = 4.02 (P < 0.0001)

Conclusions

- Incidence of symptom development in asymptomatic patients treated with a conservative approach was 23.6 %.
- Post-operative morbidity is higher when surgery is performed after symptoms develop compared to those resected while the patient is asymptomatic.
- Overall study quality was low suggesting there is a need for more high quality studies to be done in the area of CCAM management

References

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