What Is The Ideal Management Of Asymptomatic Congenital Pulmonary Airway Malformation (CPAM)?

Evidence Level III

The clinical presentation of congenital pulmonary airway malformation (CPAM) ranges from severe respiratory distress at birth to lesions which remain asymptomatic for years. An increasing proportion of neonates are presenting asymptomatically as many are now identified on antenatal ultrasound. Clinical presentation typically dictates treatment, however when cases of known cystic lung disease are asymptomatic, ideal management is less clear.

A few reports describe postnatal resolution of around 4%-13% of CPAM lesions, lending support to conservative management (Stanton 2009, Calvert 2007, Sauvat 2003, Kays 2006). However, whether this is a great enough proportion to justify observation is undetermined. It has been suggested that this phenomenon may not be true regression, but growth of healthy lung tissue around the lesion reducing its visibility on imaging (Wallis 2000).

While the natural evolution of CPAM is largely uncharacterized, it is generally assumed that persistent CPAM will eventually become symptomatic (Di Prima 2012, Fitzgerald 2007), leading to more complicated surgery and worse surgical outcomes. Therefore many authors suggest that asymptomatic patients undergo elective surgery before symptoms develop (Aspirot 2008, Conforti 2009, dell'Agnola 1996, Marshall 2000, Sueyoshi 2008, Laje 2008). In contrast, rates of symptom development have been reported as low as 3.2% indicating that surgery is unnecessary in the majority of asymptomatic patients, although this low prevalence may be attributable to inadequate follow-up length (Stanton 2009). Nevertheless, some studies have reported that there is no significant difference in post-operative outcomes between those who were symptomatic versus asymptomatic at the time of surgery, adding further support for conservative management as long as symptoms are absent (Aziz 2004, Nasr 2010).

However, the risk of malignancy, although low (1-4%; Chuang 2009), has come to play an important role in decision-making around CPAM management. Although some authors argue that the association is largely overestimated (Di Prima 2012, Fitzgerald 2007), other argue that the lethal potential of malignancy may justify surgical intervention in all patients (Priest 2009, Puligandla 2012). Additionally, over 40 cases exist detailing malignancies arising in, or being indistinguishable from, CPAM lesions lending further support to prophylactic resection of all lesions (Laberge 2005). There exist reports, however, of malignancy arising in CPAM patients after resection or in an unassociated area of the lung, suggesting that these patients may have an overall increased malignant potential which no current therapy can adequately address (Bush 2009, Chuang 2009).

The greatest argument against surgical intervention for asymptomatic CPAM lesions is the issue of unnecessarily exposing these pediatric patients to surgical risk. However, low morbidity rates and no mortalities have been associated with surgery (Tsai 2008, Eber 2007, Laje 2008), and lobectomy – the standard treatment – is well tolerated without any long term functional limitations (Beres 2011, Keijzer 2009, Naito, 2012, Nakajima 1998, Parikh 2005). It should also be noted, though, that similar functional outcomes have been reported in nonsurgically managed patients (Eber 2007), indicating that conservative management may be comparable in the long-term. Be that as it may, conservative management is associated with significantly greater exposure to radiation and anesthesia due to the use of serial CT scans for monitoring (Di Prima 2012, Eber 2007, Laje 2008). While the impact of these exposures has not been investigated, they are of concern in a pediatric population.

We identified 9 non-randomized studies from which we could extract poolable data (Aziz 2004, Chow 2007, dell'Agnola 1996, Evrard 1999, Keidar 2001, Marshall 2000, Raychaudhuri 2011, Sueyoshi 2008, Wong 2009); only one was prospective in design (Keidar 2001). One hundred sixty-eight patients were asymptomatic at birth with sufficient data available for inclusion in our analyses. Just under half of these patients (n = 70, 41.7%) underwent elective surgery with 7 (10.0 %) cases of postoperative complications. Of the remainder (n = 98, 58.3%), 63 eventually developing symptoms between one month and seven years of age and consequently required surgery (64.3%); 20 experienced postoperative complications (31.8%). The remaining 35 patients continued to be managed non-surgically, with a follow-up of between three months and nine years (35.7%). There were no deaths related to the condition or surgical complications of lung resection in any of our included patients. Meta-analysis results revealed that post-operative complications were significantly more likely when surgery was performed after symptom development compared to resection when patients were asymptomatic (OR 4.59, 95% CI 1.40 to 15.11, P = 0.01; Aziz 2004, Chow 2007, dell'Agnola 1996, Evrard 1999, Marshall 2000, Wong 2009; inf ancy, low heterogeneity (I^2 = 0%)) and that there was no significant difference in length of hospitalization after surgery (MD 4.96, 95% CI -1.75 to 11.67, P = 0.15; Marshall 2000, Sueyoshi 2008, Evrard 1999; high heterogeneity (I^2 = 63%)), although the trend favoured asymptomatic patients.

Based on these findings, we recommend elective resection of asymptomatic CPAM lesions, rather than expectant management. Lung resection in an asymptomatic patient is quite safe, presents the risk of symptom development and complicated surgery later in life, and may help prevent the development of malignancy.

Results and recommendations from non-systematic literature reviews discussing this comparison are available in this table and a summary of these results are available here.

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The full systematic review can be found here.

Non-Randomized Trials: Comparison Studies

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Non-Randomized Trials: Non-Comparison Studies

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Other Study Designs

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