

SYMPTOM DEVELOPMENT IN INITIALLY ASYMPTOMATIC CPAM DIAGNOSED PRENATALLY: A SYSTEMATIC REVIEW

Navot Kantor¹, Carolyn Wayne², Ahmed Nasr^{1,2}

¹*Faculty of Medicine, University of Ottawa, Ottawa, ON, Canada,* ²*CHEO Research Institute, Children's Hospital of Eastern Ontario, Ottawa, ON, Canada*

Aim of the study: For the approximately three-quarters of patients with a prenatal diagnosis of congenital pulmonary airway malformation (CPAM) who are asymptomatic at birth, the chance of eventually developing symptoms is unknown. Thus, we sought to explore the natural history of asymptomatic CPAM.

Methods: We searched EMBASE (1980 onwards), MEDLINE (1996 onwards), and the first 50 results from Google Scholar for literature describing the natural history of prenatally-diagnosed, initially asymptomatic CPAM, excluding editorials and case studies. Two researchers screened all studies for inclusion; one researcher extracted data and another checked it for accuracy and completeness. For asymptomatic patients managed surgically, we recorded the age at elective surgery, while for those managed conservatively, we tabulated the proportion who went on to develop symptoms as well as the mean age at symptom development.

Main results: We included data from 19 retrospective studies on 353 patients diagnosed with CPAM prenatally and born asymptomatic (103 males, 84 females, and 166 unspecified). All diagnoses were confirmed postnatally. Elective surgery was undertaken for 225 asymptomatic patients from one day to four years of age, while the remaining 128 patients were managed conservatively. Of these 128, 31 (24.2%) developed symptoms requiring surgical intervention. The mean age at symptom development, which consisted mainly of pneumonia-related complications such as fever, dyspnea, and respiratory distress, was 10 months (range: 15 days to 5 years). Curative surgery was generally performed shortly after symptom development.

Conclusion: The risk for developing respiratory symptoms exists with originally asymptomatic CPAM patients, but the exact risk is difficult to predict. Parents may be given the value of approximately 1 in 4 as an estimate of the proportion of asymptomatic CPAM patients who go on to develop symptoms, which will help them make an informed decision regarding the option of elective surgery.