

Congenital Pulmonary Malformations in Children: Diagnosis and Management in a Pediatric Department in a Low Source Country

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Dear Editor-in-Chief,

Congenital pulmonary malformations (CPM) comprise a group of anatomical abnormalities of the respiratory tree including congenital cystic malformations, bronchopulmonary sequestrations, bronchogenic cyst, bronchial atresia and congenital lobar emphysema (1). CPMs were considered rare lesions with estimates of about 1 in 10,000 to 1 in 35,000 pregnancies (2). The most common CLM are congenital pulmonary airway malformations (CPAM), pulmonary sequestration (PS), and mixed/"hybrid" lesions (3). The aim of our study was to determine the diagnosis and the management of CPM in a pediatric department in a low source country. This study retrospectively evaluated patients who had been diagnosed with CPM in the department of pediatric emergency and reanimation y in Hedi Chaker hospital in Sfax, Tunisia, between 2010 and 2016. Over 7-year period, we diagnosed and managed 8 children with CPM. Five of children were male and 3 were female. We had 2 congenital cystic malformations, 1 bronchopulmonary sequestrations, 1 bronchogenic cysts and 4 children with congenital lobar emphysema. Three of our patients had associated malformations. The prenatal diagnosis wasn't made in any of our patients. In six of our cases, symptoms appeared before the age of four months. A CT- scan has always been performed. For 6 children the treatment was surgical, and 2 patients died before being operated. Patients were operated at an average age of 11 months (21 days to 7 years). Three children maintained postoperative respiratory symptoms with a mean follow-up of 3 years. With widespread use of prenatal imaging and improving sonographic resolution, CPMs are increasingly detected in the prenatal period (1). CPMs may also present with respiratory symptoms later in childhood (1).

In our experience all CPMs were diagnosed after respiratory symptoms in age > 3 weeks. Optimal age at resection for asymptomatic CPM in the literature range from 3 weeks to <12 months (3); however, postponing surgery until patients become symptomatic results in more technically difficult resections, higher rates of post-operative complications, and poorer pulmonary function (3). In our patients the average age of operated time was 11 months and this because the diagnosis was made at a late age. The outcome in terms of mortality after CPM surgery is excellent (4). In a large series of patients operated on for CPM we found a significant prevalence of long-term morbidity as compared with controls (2). In our experience, 50% of patients operated on for CPM had clinical sequelae. Socioeconomic, demographic, and hospital factors were determinants of survival of CPM. Antenatal diagnosis should be developed in order to improve management and prognosis of CPM in country with low resource.

Key Words: Children, Congenital, Pulmonary malformations.

*Please cite this article as: Safi F, Louati H, Zghal MA, Hsairi M, Abid H, Gargouri L, et al. Congenital Pulmonary Malformations in Children: Diagnosis and Management in a Pediatric Department in a Low Source Country. *IntJPediatr*.2018;6(1):6781-82.DOI:**10.22038/ijp.2017.27786.2406**

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Received date: Dec.10, 2017; Accepted date: Dec.22, 2017

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