

# Letter to the Editor

## Surgical treatment of lung malformations in pediatric patients

Tratamento cirúrgico das malformações pulmonares em pacientes pediátricos

### To the Editor:

I was heartened to see the article on the surgical treatment of congenital lung malformations, authored by Ferreira et al. and published in a recent issue of the Brazilian Journal of Pulmonology.<sup>(1)</sup> The authors showed that patients with congenital lung malformations constitute an important group, and that the surgical procedure is safe, having provoked no deaths during the 10-year period studied. I would like to express my great admiration for the dedication of the authors to the study carried out and to the topics addressed in the article, principally the clear evidence that, in this age bracket, the surgical procedure described is a definitive and indispensable technique for the treatment of lung malformations.<sup>(2,3)</sup>

Our major difficulty lies in the accurate diagnosis of these malformations, since the clinical profile frequently varies, ranging from asymptomatic cases to cases of severe respiratory failure, in pediatric and adult patients alike.<sup>(2,3)</sup>

The simplistic term “lung malformations” does not convey the complexity of diagnosing the various diseases involved. Although central and mediastinal cysts are not located within the lung, I regard them as lung malformations.<sup>(2,3)</sup> Since the embryonic origin of these cysts is the same as that of the other congenital changes, they are bronchogenic cysts, with respiratory epithelium—pseudostratified, cylindrical and ciliated. I see no reason for studying these cysts separately from the other malformations.

Regarding the nomenclature, the term “malformation” effectively conveys the idea of congenital origin. Therefore, there is no need to employ the terms “malformation” and “congenital” in conjunction.<sup>(2,3)</sup> Can a malformation be acquired?

Cystic adenomatoid malformations, described by Stocker in 1977, are classified as one of three types (I to III), and this classification is still used.<sup>(2-4)</sup> An attempt to expand this classification, by Stocker himself, was described in 1994 and published in 2002,<sup>(5)</sup> although it has yet to be widely adopted.<sup>(4)</sup> Further studies are needed in order to show the benefits of expanding the

classification to five types. In addition, Stocker<sup>(6)</sup> proposed a change in nomenclature, from “cystic adenomatoid malformation” to “adenomatoid lung malformation”. Ferreira et al.<sup>(1)</sup> referred to this new classification as having four types (I to IV). However, other studies involving the expanded classification show five types (0 to IV), according to the origin of the malformation<sup>(4-6)</sup>: type 0 (tracheal); type I (bronchial); type II (bronchiolar); type III (bronchiolar/alveolar duct); and type IV (alveolar/distal acinar).

The scarcity of publications on this topic in Brazil reflects the difficulty in diagnosing lung malformations. Unfortunately, we treat, each year, 5–6 patients with this type of malformation at our institution,<sup>(2)</sup> similar to the numbers cited by our colleagues.<sup>(1)</sup> In the state capital of São Paulo, 64,000 patients under 9 years of age were hospitalized with pneumonia between 2004 and 2005.<sup>(7)</sup> The incidence of recurrent pneumonia (more than two episodes per year) ranges from 6% to 8%, and the cause is identified in approximately 90% of the patients.<sup>(8)</sup> Congenital pulmonary changes account for 2–9% of the cases of recurrent pneumonia.<sup>(8)</sup> Of the 64,000 children mentioned, it is likely that at least 3,840 (6%) had recurrent pneumonia. Of those, at least 76 cases of pneumonia were probably caused by malformations (2%). Therefore, there should be, in São Paulo, over 70 patients with lung malformations per year, which does not correspond to our statistics. In 1993, Adzick published a study, conducted in the USA, comprising a sample of 350 patients over a period of 7 years (approximately 50 cases/year).<sup>(9)</sup> In 2009, the same author published a study comprising a sample of over 600 patients over a period of 14 years.<sup>(10)</sup>

I report that we have treated a 14-year-old patient who had over 80 episodes of pneumonia in the right upper lobe, due to a congenital anomaly—esophageal bronchus. At this writing, the patient remains free of pulmonary infection, having experienced no recurrence since undergoing the surgical procedure in 2004. Why was this patient not correctly diagnosed earlier,

at the second or third occurrence of infection? I believe that the publication of articles on this topic is motivated by a desire to improve the diagnosis of lung malformations and to provide appropriate treatment for this condition.

We have known for several decades, since the time when they were known as “congenital lung cysts”,<sup>(11)</sup> that, prior to surgical intervention, these malformations cause complications, such as infections, hyperinflation, hemoptysis, and malignant degeneration.<sup>(11)</sup>

Ferreira et al.<sup>(1)</sup> showed that 48.5% of the patients had been treated for pneumonia before the operation, a proportion similar to the 45% that we have previously reported.<sup>(2,3)</sup> The rate of postoperative morbidity (complications) reported by the authors was 28.5%,<sup>(1)</sup> compared with 23% in our studies.<sup>(2,3)</sup> It is evident that the proportion of complications is almost twice as high when we opt for “monitoring” rather than surgical treatment in the management of lung malformations. Therefore, I have no doubts that, even in asymptomatic pediatric patients, resection of lung malformations is indicated as soon as the diagnosis has been made. In addition, treating patients with lung malformations early (before they reach 8 years of age) allows compensatory lung growth, with no loss of pulmonary function.<sup>(1,2,4)</sup> In asymptomatic cases, I recommend that the surgical procedure not be employed in patients who weigh less than 10 kg or are younger than 12 months of age. In patients above 12 months of age, we have observed no mortality.<sup>(2,3)</sup>

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