Journal of Pediatric Surgery 52 (2017) 1891–1897

Contents lists available at ScienceDirect

Journal of Pediatric Surgery

journal homepage: www.elsevier.com/locate/jpedsurg

Clinical

Long-term lung function in children following lobectomy for congenital lung malformation

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ARTICLE INFO

Article history:
Received 7 August 2017
Accepted 28 August 2017

Key words:
Congenital lung malformation
Lobectomy
Pulmonary function tests
Children
Long-term outcome

ABSTRACT

Background/purpose: To date, the optimal management of asymptomatic congenital lung malformations (CLMs) is still debated. There is still scant and controversial information regarding the long-term assessment of pulmonary function (PF) after lobectomy in children. The aim of this study is to evaluate PF in children who underwent lobectomy for CLM in infancy, hypothesizing that patients operated during the first year of life retain a normal lung function.

Methods: Children operated between 2005 and 2016 at our institution underwent PF evaluation through spirometry/whole-body plethysmography, forced oscillation technique, and multiple-breath inert gas wash-out.

Results: Out of 85 patients who underwent lobectomy at a median age of 5 months, 50 met the inclusion criteria, and 28 patients were tested. More than 80% of patients had normal FEV_1 and FVC. The mean FEV_1, FVC, $FEF_{25–75\%}$ values were higher in the patients operated before reaching one year of age.

Conclusions: The long-term outcome after lobectomy was excellent for most patients, as they retained a normal long-term PF. Therefore, for asymptomatic patients, a surgical approach before one year of age to avoid complications such as malignancy and to ensure an optimal PF catch-up could be beneficial.

Level of evidence: III – Treatment Studies.

Congenital lung malformations (CLMs) comprise a broad spectrum of rare anomalies, some of which result from a focal developmental abnormality, such as congenital pulmonary airway malformation (CPAM), extra- and intra-lobar bronchopulmonary sequestration (BPS), congenital lobar and segmental emphysemas (CLE), together with less common diseases such as bronchogenic cyst and bronchial atresia [1–3]. CPAM – previously known as congenital cystic adenomatoid malformation (CCAM) – is an intra-pulmonary benign mass of non-functioning pulmonary tissue that is usually confined to one lung lobe, although a multi-lobe involvement has been rarely observed [4]. BPS is a non-functioning solid lung mass characterized by a systemic arterial supply and by a venous drainage that may occur through the pulmonary vein. CLE, on the other hand, is a rare anomaly due to the overinflation and distension of one or more pulmonary lobes, which usually occurs on the upper left or right middle lobe [1,5]. The reported incidence of CLM is between 1 in 10,000 to 1 in 35,000 live births, with no predilection for lung side, sex or race [4,6–8], although true incidence is yet to be determined due to the increasing rate of prenatally diagnosed lesions. The natural history of prenatal CLMs varies from complete regression in utero to life-threatening fetal hydrops. In case of fetal hydrops, fetal surgery is nowadays considered a therapeutic option if there has not been a significant response to maternal betamethasone therapy [4,6]. Prenatally diagnosed CLMs may regress at antenatal scans in up to 15% of the cases. The timing of regression is variable and usually occurs during the mid-third trimester at 32–34 weeks of gestation. The newborn might present itself at birth with significant cardiorespiratory compromise. Conversely, in other cases the patients may be asymptomatic at birth and the lung lesion may be incidentally diagnosed during postnatal life [3,4]. While there is a consensus on symptomatic congenital lung lesions treatment, the optimal management of asymptomatic lesions remains controversial: both timing and the need for surgical

Abbreviations: CLM, congenital lung malformation; PF, pulmonary function; CPAM, congenital pulmonary airway malformation; BPS, bronchopulmonary sequestration; CLE, congenital lobar emphysema; FOT, forced oscillation technique; FVC, forced vital capacity; FEV_1, forced expiratory volume in 1st sec; $FEF_{25–75\%}$, forced expiratory flow rate between 25 and 75% of vital capacity; TLC, total lung capacity; RV, residual volume; FRC, functional residual capacity; $LCI_{5\%}$, lung clearance index 5%; $LCI_{2,5\%}$, lung clearance index 2,5%; R(8), average resistance at 8 Hz; X(8), average reactance at 8 Hz.

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http://dx.doi.org/10.1016/j.jpedsurg.2017.08.059
0022-3468/

excision remain an ongoing topic of debate. Some centers prefer early elective resection of these lesions, before the onset of symptoms related to complications such as recurrent pulmonary infection, pneumothorax and malignancy. This operative approach is also supported by the concept of compensatory growth of the remaining lung following surgery, which could be enhanced by a lobectomy performed during early infancy [9–14]. On the other hand, the limited data documenting the long-term pulmonary morbidity resulting from lung resections in infancy, led other authors to prefer an active observation until patients became symptomatic [7,15,16]. Few studies about pulmonary function (PF) after lung resection in infancy are available, with incomplete and non-exhaustive PF evaluation and small sample series [17–26]. In the following study, we report the experience of a single institution on CLMs in children who underwent lobectomy over a ten-year period. With the aim to gain consensus on the operative management of asymptomatic lung lesions, we hypothesized that most of the patients who underwent lobectomy had a long-term PF within the normal range, since most of them were operated during the first year of life.

1. Methods

This study received IRB approval (No. 147/2016). Medical records were retrieved for all those children who underwent lobectomy for benign CLM between January 2005 and January 2016 at Meyer Children Hospital. The following data were extrapolated: number of patients, date of birth, sex, age at surgery, type of disease, type of operation, number of operations, comorbidities, address and telephone number. Patients were considered for this study if they met the inclusion criteria as indexed below:

Inclusion criteria:

1. Any patient who underwent previous lobectomy for CLM at Meyer Children Hospital.
2. Any patient who was 3 years of age or older, at the time of PF tests.

Exclusion criteria:

1. Any patient younger than 3 years of age, at the time of PF tests.
2. Any patient who underwent any other thoracic surgical procedure (e.g. pneumonectomy, bi-lobectomy, extralobar BPS excision)
3. Deceased patients.
4. Presence of comorbidities: diffuse lung disease (e.g. cystic fibrosis), bronchomalacia, tracheomalacia, laryngomalacia, neurologically impaired patients, severe preterm babies, syndromic patients.

As described in the flow-chart (Fig. 1), a total of 85 patients (51 male and 34 female) with CLM were identified at our institution during the study period. All of them underwent lobectomy for CLMs (CPAM, BPS, CLE or hybrid lesion). A brief information note was handed to the families of those patients who met the inclusion criteria. Subsequently, as shown in Fig. 1, albeit some attempts were unsuccessful, most of the parents were contacted by phone call. After several attempts to contact the patients' family, only 32 of them accepted to participate in our study. Of the 32: three patients, all aged 3, failed the tests after several attempts for lack of cooperation and one patient did not show up at the time of the tests. In conclusion, 28 patients were assessed at our Pulmonary Unit (17 males and 11 females).

Patients which were enrolled, whose parents or legal guardians consented to this study, underwent physical examination and PF evaluation through the following three techniques: spirometry/whole-body plethysmography, forced oscillation technique (FOT), and multiple-breath nitrogen wash-out in 100% oxygen. Measured or estimated parameters included forced vital capacity (FVC), forced expiratory volume in first second (FEV_1), forced expiratory flow rate between 25 and 75% of vital capacity ($FEF_{25-75\%}$), total lung capacity (TLC), residual volume (RV), RT/TLC ratio, functional residual capacity (FRC), lung clearance index 5% ($LCI_{5\%}$) and lung clearance index 2,5%($LCI_{2,5\%}$), average resistance at 8 Hz R(8) and average reactance at 8 Hz X(8). Multiple-breath nitrogen wash-out measurements were useful to assess

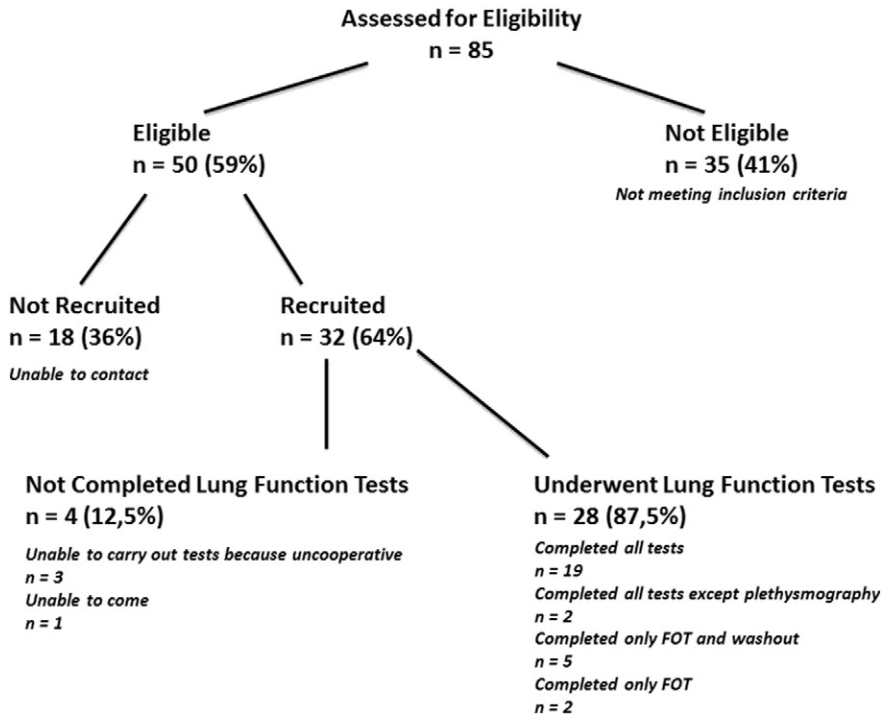

Fig. 1. Enrolment flow chart. Eighty-five patients were identified, all of them underwent lobectomy for CLM; 50 patients met inclusion criteria and 32 of them were recruited and accepted to participate to our study; 28 patients were tested and 4 children not completed lung function test; 21 of 28 patients completed all three tests, although plethysmography data was unavailable in two cases; 5 children completed only FOT and wash-out test, and 2 children only FOT.