Case Series

Is surgical intervention routinely required for congenital lobar overinflation? A case series from a tertiary hospital in Riyadh, Saudi Arabia

Hanan I. AlOmran\textsuperscript{a,}\textsuperscript{*}, Ibrahim AlMogarri\textsuperscript{b}, Sami AlHaider\textsuperscript{b}, Mohammed AlZaid\textsuperscript{c}, Khaled AlThobaiti\textsuperscript{d}

\textsuperscript{a} Pediatric Department, College of Medicine, Prince Sattam Bin Abdulaziz University, AlKhurj, Saudi Arabia
\textsuperscript{b} Pediatric Department, King Fahad Specialist Hospital and Research Center (KFSHRC), Riyadh, Saudi Arabia
\textsuperscript{c} Pediatric Department, King Fahd Medical City (KFMC), Riyadh, Saudi Arabia
\textsuperscript{d} Pediatric Department, Taif Children Hospital, Taif, Saudi Arabia

\textbf{A B S T R A C T}

Introduction: Congenital lobar overinflation (CLO) is a congenital overinflation of a pulmonary lobe. The treatment choice depends on the severity of its symptoms. Surgical intervention is indicated for patients with significant symptomatology, while a conservative approach is used to treat incidental and mildly symptomatic lesions. However, the conservative approach for children with mild symptoms is not very common among pulmonologist. Therefore, we evaluated this approach to treating mildly symptomatic children.

Methods: This retrospective study examined mildly symptomatic patients (n = 14) with a radiological diagnosis of CLO between June 2005 and August 2018 who were treated conservatively at KFSHRC in Riyadh. The participants’ ages ranged between two days and four years, with follow-up period ranged from four months to 10 years.

Results: Fourteen patients with CLO—who were 2 days to four years old and comprised 10 boys (71.4%) and four girls (28.6%)—were treated conservatively. All patients were symptomatic upon presentation, and their main clinical findings were tachypnea (85.7%) and dyspnea (78.6%). A single lobe was affected for ten patients (71.4%). Congenital cardiac anomalies founded in six patients (42.9%). Radiological image showed overinflation of all patients’ affected lobes. Significant mediastinal displacement was observed among two patients (14.3%). During their follow-up periods, nine patients (64.3%) became asymptomatic, three (21.4%) showed improvement, and two (14.3%) remained symptomatic and underwent lobectomy.

Conclusions: The good outcomes for mildly symptomatic children with CLO in our series indicate that the conservative approach can be considered to treat these children at any age, along with close follow-up.

1. Introduction

Congenital lobar Overinflation (CLO), previously known as congenital lobar emphysema, is a developmental lung malformation characterized by hyperinflation of the partial or entire pulmonary lobe [1]. CLO’s cause is unknown in 50% of cases [2]. Incomplete or defective bronchial cartilage, causing the bronchi to collapse upon expiration and obstruct deflection, is the most commonly identified cause of CLO, occurring in 25% of cases and resulting in airway obstruction [3–5]. Airway obstruction can result from an internal bronchial obstruction or an external bronchial obstruction, though internal obstructions are more common [2]. Internal bronchial obstructions are caused by bronchial stenosis or atresia, bronchomalacia, an endobronchial obstruction from mucosal proliferation, or abnormal bronchi [3,4,6]. Meanwhile, abnormal cardiopulmonary vasculature—such as a pulmonary arterial sling anomaly, a pulmonary rotation anomaly, bronchogenic cysts, duplication of the esophagus, and mediastinal cysts or tumors—have been found to cause external bronchial obstructions [3,4,7].

The clinical presentations are variable. This malformation may present with acute respiratory failure among newborns, recurrent chest infections, recurrent episodes of tachypnea, or chronic cough [8,9]. A minority of children with CLO remain asymptomatic [10]. Surgical resection or lobectomy is generally considered the treatment of choice for severe CLO cases [11]. While a conservative approach is used to treat...
asymptomatic and mildly symptomatic newborns and children with CLO [12,13].

Accordingly, in our study, we retrospectively evaluated 14 mildly symptomatic patients with CLO who had been treated conservatively. We also describe their long-term follow-up results and compare our findings with other cases reported in the literature.

2. Methods

A case series study, we retrospectively reviewed the charts of 14 patients who had been diagnosed radiologically with CLO between June 2005 and August 2018 and treated conservatively at King Faisal Specialist Hospital and Research Center (KFSHRC) in Riyadh, which is the largest tertiary care center in Saudi Arabia and accept referral from all Saudi Arabia and Gulf Club Countries (GCC).

Participants’ age upon their initial presentation to our hospital ranged between two days to four years (Diagram 1).

Patients’ records were evaluated regarding their age, gender, clinical presentations, radiological findings at the time of their diagnosis and follow-up appointments, associated diseases, and outcomes. Their mean follow-up duration was 53 months (range: 4 months to 10 years).

Descriptive statistics were used to summarize our data. The results are expressed, as appropriate, as percentages, means, and ranges. This study received ethical approval from the King Faisal Specialist Hospital Research Ethics Committee.

This study is registered at Chinese clinical trial registry. Unique Identifying number (ChiCTR2000055634). Also, it is reported according to STROCSS criteria [14].

3. Results

The 14 children who had presented to KFSHRC between June 2005 and August 2018 with radiological findings suggesting CLO, and who were treated conservatively, are represented in Diagram 1. Patients’ clinical and radiological findings at the time of their diagnosis are summarized in Table 1.

Participants’ mean age was 11 months (range: two days to four years). Ten patients were male (71.4%) while four patients were female (28.6%). A single lobe was affected for 10 patients (71.4%), of whom the left upper lobe was affected for six patients (42.9%), versus the right upper lobe for two patients (14.3%) and the right middle lobe for two patients (14.3%). Meanwhile, for four patients (28.6%), two lobes were affected. All of this study’s patients were symptomatic upon their initial presentation to our hospital, and their main symptoms and clinical findings were tachypnea (12 patients; 85.7%), dyspnea (11 patients; 78.6%), recurrent chest infection (six patients; 42.9%), and cough (three patients; 21.4%). Oxygen saturation levels were in the normal range (94% and higher, as measured by pulse oximeter) for 12 patients (85.7%), while desaturation was detected for two patients (14.3%), both of whom had congenital heart disease. Participants’ growth parameters upon their presentation were within normal limits for 13 patients (92.9%). One patient (7.1%), however, was found to exhibit a failure to thrive, and he had complex congenital heart disease.

Congenital heart anomalies were detected among six patients (42.9%)—including patent foramen oval, patent ductus arteriosus, a ventricular septal defect, an atrial septal defect, pulmonary hypertension, a small left pulmonary artery, pulmonary atresia, transposition of the great artery, and tetralogy of Fallot). One patient was found to have hypermobile joint syndrome (7.1%).

Conventional chest radiographs showed overinflation in all cases, and atelectasis of the ipsilateral or contralateral lobe or lobes was present in 11 cases (78.6%) (Fig. 1). Significant mediastinal displacement with herniation of the affected lobe across the mediastinum was observed in two cases (14.3%). Chest computerized tomography (CT) was performed for all patients, showing overinflation and vascular attenuation of the all affected lobe or lobes, atelectasis of the ipsilateral or contralateral lobe or lobes in 12 cases (85.7%), and significant mediastinal displacement with herniation of the affected lobe across the mediastinum in two cases (14.3%). Both of these two latter cases were failed conservative approach (Fig. 2).

Patients’ clinical and radiological findings during their follow-up period are summarized in Table 2. The mean follow-up duration for all patients was 53 months (range: four months to 10 years). During the follow-up periods of all 14 patients, nine patients (64.3%) became asymptomatic, three patients (21.4%) showed clinical improvement, and two patients (14.3%) remained symptomatic, both of whom underwent a lobectomy at five and 23 months of age. All patients’ oxygen saturation levels were in the normal range (94% and above), as measured using a pulse oximeter. Moreover, 12 patients’ growth parameters during the follow-up period were within the normal limits (85.7%). However, two patients (14.3%) were found to exhibit a failure to thrive, and they both underwent lobectomy.

Follow-up imaging was performed for all patients. No significant

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Table 1

| Parameter          | N = 14 |
|--------------------|--------|
| **Age at diagnosis** | 2 days–4 years |
| **Gender**         | Male 10 (71.4%); Female 4 (28.6%) |
| **Types of CLO**   | Single 10 (71.4%); Multilobe 4 (28.6%) |
| **Location**       | LUL in 6 (42.9%); RML in 2 (14.3%); RUL in 2 (14.3%); LUL + RML in 1 (7.1%); RUL + LUL in 2 (14.3%); RML + RLL in 1 (7.1%) |
| **Association**    | Isolated in 7 (50%); Associated with CHD in 6 (42.9%); Associated with hypermobility joint syndrome in 1 (7.1%) |
| **Sign and symptoms** | Tachypnea in 12 (85.7%); Dyspnea in 11 (78.6%); Cough in 3 (21.4%); Recurrent chest infection in 6 (42.9%); Desaturation (<94%) in 2 (14.3%) |
| **Growth parameter** | Within normal limit in 13 (92.9%); FFT 1 in (7.1%) |
| **CXR**            | Hyperlucency in 14 (100%); Atelectasis in 11 (78.6%); Significant mediastinal shift with lobar herniation to opposite side in 2 (14.3%) |
| **CT scan**        | Overinflation in 14 (100%); Atelectasis in 12 (85.7%); Shift to opposite side in 2 (14.3%) |

LUL left upper lobe; RML, right middle lobe; RUL, right upper lobe; CHD, congenital heart disease; FTT, failure to thrive.

*FTT if weight less than 3rd percentile or cross two major percentile in CDC growth chart.
Radiological changes were found for five patients (35.7%), while six patients (42.9%) demonstrated radiological improvements (reduced overinflation), and one patient had resolution of radiological findings (7.1%). However, two patients (14.3%) demonstrated radiological worsening (overinflation and significant mediastinal shifting) (Fig. 3).

Pulmonary function tests were performed for three patients. There tests’ results were normal for one patient, and obstruction patterns were detected for two patients. No patient lost the follow-up.

4. Discussion

CLO is a developmental lung malformation characterized by hyperinflation of one or more of the pulmonary lobes [1]. CLO’s cause is unknown in 50% of cases, and its exact cause is difficult to identify [2]. However, the most identified cause of CLO is an obstruction of the developing airway, which occurs in 25% of cases. Male patients appear to be affected by CLO more than female patients [3,4], and our findings were similar to the literature’s findings in this regard since 71.4% of our patients were male. CLO is usually unilateral, most commonly affecting the left upper lobe, followed by the right middle lobe and right upper lobe, although bilateral involvement has also been reported [15]. The lobe distribution in our series was consistent with the literature’s distribution, with uni-lobar involvement most commonly affecting the left upper lobe, followed by the right middle lobe and right upper lobe, more commonly than multi-lobar distribution. However, in cases of multi-lobar involvement, we found that the right upper and left upper lobes were commonly affected.

A large number of concomitant malformations accompany CLO, particularly cardiovascular malformations [1]. In our series, seven patients (50%) were found to have associated abnormalities. Congenital heart defects were found in six cases (42.9%), and hypermobile joint syndrome was found in one case (7.1%). CLO presentations are variable in terms of patients’ ages and symptoms. Previous studies have found that the condition is present at birth for approximately 25–33% of patients, manifesting at one month of age for 50% of patients and present among nearly all patients by six months of age [2,16]. Patients’ age at the time of their CLO diagnosis is closely related to the severity of their respiratory distress [1]. In our series, patients’ ages at the time of their diagnosis ranged from two days to four years, with a mean of 11 months, which is older than the mean reported in the literature. The reason for this difference is the presence of milder respiratory symptoms among our study’s patients, which resulted in a need to seek medical help later. Most of our patients presented with mild respiratory distress. The main symptoms and clinical findings upon their initial presentation to our hospital were tachypnea, dyspnea, recurrent chest infection, and cough. Desaturation (oxygen saturation of less than 94% as measured by pulse oximeter) was present among two patients (14.3%), both of whom had congenital heart disease.

CLO is often diagnosed based on its characteristic appearance in chest radiographs [17]. Chest film typically shows overinflation of the affected lobe with indistinct vessel markings, atelectasis of the ipsilateral and contralateral lung lobes, a mediastinal shift, and herniation of the affected lobe to the opposite side [1]. In our series, overinflation of the affected lobe was observed for all patients, while atelectasis at the
ipsilateral or contralateral lobe or lobes was observed among 11 patients (78.6%). A significant mediastinal shift with herniation of the affected lobe to the opposite side was observed in two cases (14.3%). Chest CT showed the anatomy of patients’ airways down to the segmental level, allowing a visualization of the intrinsic or extrinsic sources of airway obstruction [11]. Chest CT was performed for all patients in this study, showing similar findings to the chest radiographs in more radiological details.

CLO treatment varies, based on patients’ clinical severity. While lobectomy is recommended for clinically progressive and severely symptomatic patients, and conservative management is an option for asymptomatic patients, the treatment approach for children with mild symptoms is less clear [8]. Tharkal et al. [9] reported on seven mildly symptomatic patients who were treated conservatively, finding that such treatment failed for almost half of these patients, who then underwent surgery. Meanwhile, other studies have reported on a small proportion of children treated conservatively with good outcomes [10, 11]. In our series, all patients were treated conservatively, and during the follow-up period, most patients showed clinical and radiological improvements. Of our 14 symptomatic participants, 12 patients were

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**Diagram 1.** Flowchart of study design.

Suspected congenital lobar overinflation (CLO)

1. Lung lobe over-inflation secondary to other pathological causes (cystic lung malformations, foreign body aspiration)
2. Significant respiratory distress
3. Chest asymmetry

Confirm CLO radiologically with mild symptom (n=14)

**Chest radiograph & CT**

Overinflation
Atelectasis of neighbor lobe
Significant mediastinal displacement with herniation of the affected lobe across the mediastinum (n=2)

Outcome
Lobectomy (failed conservative management) in 2 (14.3%)

**Chest radiograph & CT**

Overinflation
Atelectasis of neighbor lobe (n=12)

Outcome
Conservative management in 12 (85.7%)

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conservatively treated successfully—nine patients (64.3%) became completely asymptomatic, and three patients (21.4%) showed clinical improvements. Meanwhile, two patients (14.3%) remained symptomatic and underwent lobectomy at five and 23 months of age. (Table 2). Most of our participants showed radiological improvements, and one achieved complete resolution of radiological abnormality.

Karnak et al. [11] recommend surgery to treat CLO among all infants younger than two months and a conservative approach for mildly to moderately symptomatic patients older than two months. Mei-Zahav et al. [8] recommend conservative treatment for mildly symptomatic children with CLO at any age. Based on our data, patients’ clinical and radiological improvements during their follow-up periods support a recommendation for the conservative treatment of mildly symptomatic CLO patients. Once a conservative treatment has been chosen, patients should be closely observed for any indication of a possible future lobectomy, according to their clinical situation.

5. Conclusion

In our series, the good treatment outcomes for children with mildly symptomatic CLO indicate that conservative treatment approaches can be considered for such children at any age, along with close follow-up for any indication of a possible future lobectomy, according to their clinical situation. However, patients with CLO who present with clinical findings of severe respiratory distress, severe hypoxemia, hypercarbia, a failure to thrive, or a significant mediastinal shift with herniation of the affected lobe across the mediastinum should be operated upon as soon as possible.

This study involved a large group of patients, evaluating the use of conservative treatments for mildly symptomatic children with CLO. However, more research needed with multicenter collaboration to ensure a large number of participants and to improve generalizability of the results.

Limitations

This study only focused on patients who had been treated conservatively, and this approach does not allow a histological diagnosis. Therefore, the CLO diagnoses in this study were based on imaging. Patients with other pathologies that show similar radiographic changes to CLO might have been included in this and other series.

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Ethical approval

This study was ethically approved by King Faisal Specialist Hospital Research Ethics Committee. REF: C380/203/40.

Consent

The patients’ parents have provided permission to publish these features of their children cases, and the identity of the patients have been protected.

Author contribution

IM designed the study, conducted the research, provided research materials, supervising the project, and editing the manuscript. SH provided research materials, HO participated in the coordination of the study, collected, analyzed the data, and writing the manuscript. MZ wrote the proposal, collected and entered the data. KT helped in literature review and collected the data. All authors have reviewed and approved the final draft.

Registration of research studies

1. Name of the registry: Chinese clinical trial registry
2. Unique Identifying number or registration ID: ChiCTR2000055634
3. Hyperlink to your specific registration (must be publicly accessible and will be checked): Link to research click here

Guarantor

Hanan I AlOmran. Consultant Pediatrician and Pediatric Pulmonologist +966567334563 h.alomran@psau.edu.sa.

Declaration of competing interest

The authors have no conflicts of interest to declare.

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Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.jamsu.2022.103409.

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