Review Article

Comparative Analysis of Surgical Strategies in Bilateral Congenital Lobar Emphysema in Children: A Systematic Review

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Abstract

Introduction: Bilateral congenital lobar emphysema (BCLE) is a rare developmental anomaly characterised by overinflation of one or more pulmonary lobes, leading to respiratory distress in neonates and infants. Though the management of unilateral or bi-lobar congenital lobar emphysema (CLE) is straightforward, lobectomy is the standard management. Management strategies for BCLE are often not clear and straightforward. This systematic review aims to evaluate the existing literature regarding the management of BCLE, including surgical and non-surgical approaches. **Methods:** A systematic search of electronic databases, including PubMed, SCOPUS, EMBASE and Cochrane Library, was conducted for articles published up to January 2024. The search strategy included keywords such as 'bilateral congenital lobar emphysema', 'management', 'treatment', 'surgery', 'medical management', 'bilateral CLE' and their variations using Boolean operators like AND OR and NOT. We included all the studies that reported on management strategies, outcomes and complications of BCLE in children. All studies with duplicate records across the databases, deemed irrelevant to the topic of interest and with no available full text and/or details of the intervention or its outcome were excluded. We also excluded a case report involving an adult with BCLE. Data extraction and synthesis for this review were performed following the Preferred Reporting Items for Systematic Reviews and Meta-Analysis 2020 guidelines. **Results:** The initial search identified 63 articles, of which 15 met the inclusion criteria after all the screening; the included studies comprised case reports, case series, retrospective cohort studies and systematic reviews. Twenty-one children were reported across these studies, and they were either managed conservatively or underwent sequential or simultaneous surgery. Four deaths occurred: two occurred prior to the planning of any intervention, one occurred after simultaneous surgery, and another occurred prior to the surgery on the contralateral side. The majority (n = 12, 75%) of the cases were managed by sequential surgery in the literature. **Discussion:** The management of BCLE remains challenging, with no consensus regarding the optimal approach. Surgical resection, particularly lobectomy, is often necessary to alleviate symptoms and prevent complications. However, the timing and extent of surgery require individualised consideration based on the severity of symptoms and radiological findings. We attempted to analyse the surgical strategies employed in managing these cases and their results, and we attempted to ascertain the optimal management strategy based on the review. **Conclusion:** This systematic review highlights the diverse management strategies employed in treating BCLE. Surgical resection remains the cornerstone of therapy, although there is no clarity regarding the timing and extent of surgery and the approach varies among clinicians and centres. Non-surgical approaches may have a role in select cases.

Keywords: Bilateral, congenital lobar emphysema, congenital lobar emphysema, surgery

Introduction

Bilateral congenital lobar emphysema (BCLE) is a rare developmental anomaly characterised by overinflation of one or more pulmonary lobes, leading to respiratory distress presenting either at birth itself or early infancy. Although the management of unilateral congenital lobar emphysema (CLE) is straightforward with lobectomy being the standard management. Management strategies for BCLE are often not clear and straightforward. Surgical management is usually required in symptomatic cases, and paediatric surgeons are faced with the dilemma of whether to resect both lobes simultaneously or sequentially. Proponents of the former approach argue that with advances in paediatric anaesthesia and post-operative intensive care management, it is possible

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to perform bilateral lobar resections in infants. The others favour a sequential route with the rationale that infants tolerate a unilateral lobectomy better and, in some cases, patients might not even warrant a contralateral lobectomy.

This systematic review has been done to describe the patient demographics, modes of presentation, pre-operative, intraoperative and post-operative management and the outcomes of various management strategies for BCLE. The aim of the study was to conduct a systematic review on the various management strategies for children with bilateral CLE and to study their outcomes and to arrive at a consensus as per available literature for optimal surgical approach in BCLE.

Methods

Literature search

A systematic review was conducted on the management of BCLE in children by adhering to the Preferred Reporting Items for Systematic Reviews and Meta-analysis (PRISMA) guidelines, 2020,[1] at the Department of Paediatric Surgery, All India Institute of Medical Sciences, Jodhpur, India, from March 2024 to April 2024.

An extensive search across PubMed, SCOPUS, EMBASE and Cochrane Library was done. The strategy employed was the use of search terms with Boolean operators like 'bilateral AND lobar AND congenital AND emphysema' OR 'bilateral AND CLE'. Duplicate records that appeared irrelevant to the topic of interest and without a full text available for analysis were excluded. The remaining studies were reviewed in detail after retrieval of the full version of the article. Studies in adults with BCLE were excluded [Figure 1].

Study selection and data extraction

Titles and abstracts were screened by two independent authors (SK, AS) for eligibility, in accordance with the PRISMA 2020 checklist. All studies that had management of 'bilateral congenital lobar emphysema' or 'bilateral CLE' in children up to January 2024 were included. The studies were grouped as individual case reports, case series, retrospective cohort studies and previously done systematic reviews. Initial database search identified 63 relevant papers. After duplicate studies and studies irrelevant to our study were removed, 20 records were reviewed. Four articles did not have full text available due to very old publication dates, leaving 16 abstracts available for screening. Full-text articles were systematically reviewed and further re-assessed by additional authors (SK, MP, KR and RS) for suitability. Once the additional authors approved the studies, data from all the studies were retrieved by one author (SK) into Microsoft Excel. All cases from the above studies were sequentially numbered in chronological order of publication. The data entry itself was doubly reviewed and confirmed by the other author (AS).

Data items

Data retrieved from each case report included demographic data like age, gender and country of origin, clinical data like mode of presentation, associated cardiac anomalies, initial diagnosis, pre-operative investigations (Chest X-ray [CXR], computed tomography [CT] scan, bronchoscopy and radionuclide imaging), the lobes involved, type of intervention whether operative or non-operative, simultaneous versus sequential surgery and their rationale, anaesthetic and surgical challenges, placement and number of intercostal drains (ICDs), duration of ICD, duration of post-operative stay outcome and follow up. Data items with missing information/unclear information in the case descriptions were treated as 'Not available (NA)' and excluded from the statistical analysis.

Study risk of bias and quality assessment

The methodological quality of case series studies (n = 4) and case reports (n = 11) was done using the Joanna Briggs Institute's (JBI) critical appraisal tools for case series and case reports.[2,3] A 4-point marking was done for each of the domains, and a colour coding scheme was used to represent this information: 'Yes' – Green, 'No' – Red, 'Unclear' – Yellow and 'Inappropriate' – Black.

Effect measures and methods for synthesis

The effect measures used for presentation of results were median, mean and percentages. Tables were used to describe the demographic data (that includes country of origin, year of publication, age at symptom onset, age at surgery and gender) of cases included in all studies. Appropriate filters were used to eliminate data items marked 'NA' analysis of the data. A flow diagram is used to summarise the outcomes of the broad treatment approaches to BCLE – Conservative (medical) management, sequential surgery and simultaneous surgery.

Results

Study selection

The study was commenced by identifying the area of interest as management of BCLE in children and performing a literature search across the databases SCOPUS, PubMed, EMBASE and Cochrane Library. A total of 63 records have been identified across all databases. Before the screening, duplicate records (n = 22) and records deemed irrelevant to the topic of interest (n = 21) have been removed. The remaining records (n = 20) were found, of which 4 of them were eliminated as there were no available full texts for the analysis of the kind of intervention performed and the outcomes. The retrieval of complete records was done for the remaining (n = 16), of which one was eliminated as it was found to contain a case report of a BCLE in an adult. Thus, 15 studies were considered for final analysis and interpretation which comprised of 21 patients [Figure 1]

Study characteristics

A total of 15 studies were found, of which 11 were single case reports, while four of them were authored by Garge *et al*.,[4] Perea *et al*.,[5] Stigers *et al*.[6] and Ghribi *et al*.[7] were case series. Of these, Ghribi *et al*.[7] described a series of three cases of bilobar CLE, and only one of them was a bilateral CLE;

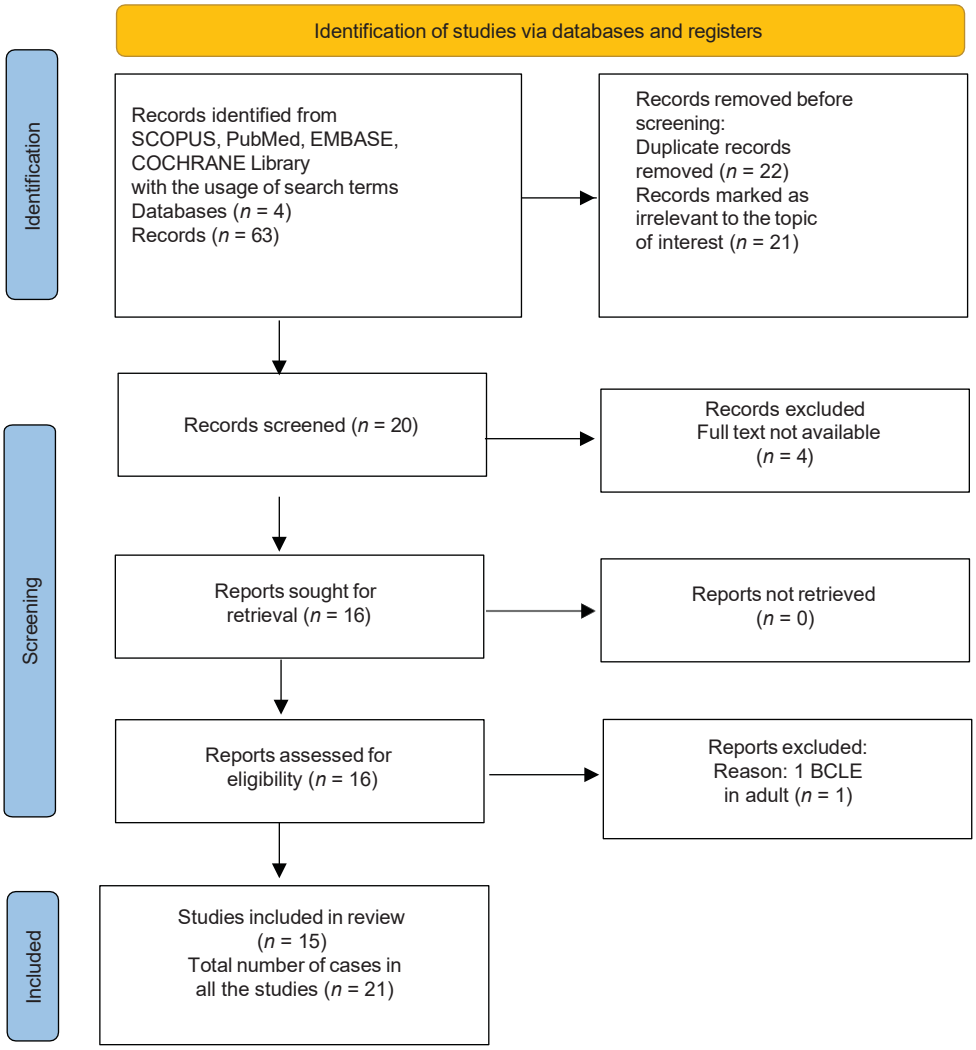


Figure 1: Preferred Reporting Items for Systematic Reviews and Meta-analysis diagram for our systematic review

only one case was taken into consideration. This amounted to a total of 21 reported cases of BCLE.

Quality and risk of bias assessment

The studies were assessed with the utilisation of the JBIs critical appraisal tools for case series and case reports. Although all of the included studies had an inherent bias given the descriptive nature, they were well constructed and have scored well as far as the methodological quality was concerned. Thus, all the case series ($n = 4$) and case reports ($n = 11$) were included in the review [Figures 2 and 3].

Study results

Data retrieved included demographic data, clinical and operative data, post-operative and follow-up data. Demographic data, including year of publication, country of origin, age of onset of symptoms, age at surgery and gender, have been collected and represented in a tabular format [Table 1]. The majority of the cases reported were from the USA ($n = 7$), followed by India ($n = 6$), and the rest of the cases were single reports from 8 different countries, as enlisted in the above table.

Age at presentation

Age at presentation ranged from birth to 2.5 months, with the majority of cases, $n = 12$ (57.14%) ≤10 days of postnatal life, $n = 7$ (33.33%) ≤1 month of life and $n = 2$ (9.5%) ≥2 months of life. Age at surgical intervention ranged from 15 days to 210 days of life (Mean: 91.5 days). Male: female ratio was 3:1.

Preoperative investigations

All cases across studies had their diagnosis made after a preoperative CXR and CT scan. A radionuclide V/Q scan was obtained in 3 out of 21 cases (14.28%), and an intraoperative bronchoscopy was done in 10 out of 21 (47.61%) cases. Two-dimensional echocardiography to rule out associated cardiac anomalies was done in 7 out of 21 cases (33.33%)– the findings of which were normal in three ($n = 3$); atrial septal defect in one ($n = 1$), patent foramen ovale in two ($n = 2$) and a right ventricular hypertrophy in one child ($n = 1$).

The initial diagnosis in 8 (39.1%) cases was suggestive of BCLE, and these cases presented earlier in the neonatal age group. In 1 (61.9%) cases, the presentation was not typical of CLE and

Table 1: Demographic data of bilateral congenital lobar emphysema cases across existing published literature

Author	Year	Country	Age at symptom onset	Age at presentation in days	Gender (male/female)*
Schiller *et al*.[8]	1970	USA	Birth	75	Female
Ekkelkamp and Vos[17]	1987	Netherlands	Birth	NA*	Male
Stigers *et al*.[6]	1992	USA	2.5 months	75	Male
Stigers *et al*.[6]	1992	USA	Birth	60	Female
Maiya *et al*.[14]	2005	UK	1 day	30	Male
Kumar *et al*.[10]	2006	India	1 month	180	Male
Ghribi *et al*.[7]	2008	Tunisia	1 month	45	Male
Iodice *et al*.[12]	2008	London	1 week	30	Male
Abushahin *et al*.[15]	2012	Qatar	1 month	60	Male
Perea *et al*.[5]	2017	USA	2 days	180	Female
Perea *et al*.[5]	2017	USA	10 days	18	Male
Perea *et al*.[5]	2017	USA	Birth	15	Male
Perea *et al*.[5]	2017	USA	2 months	210	Male
Lei *et al*.[11]	2020	China	6 days	75	Male
M A *et al*.[9]	2020	Morocco	1 month	90	Male
Sawant *et al*.[16]	2021	India	1 month	120	Male
Garge *et al*.[4]	2022	India	Birth	180	Male
Garge *et al*.[4]	2022	India	1 month	210	Female
Garge *et al*.[4]	2022	India	Birth	120	Male
Zinaye and Zeray[18]	2022	Ethiopia	6 h	30	Female
Duraiswamy *et al*.[13]	2023	India	23 days	27	Male

*NA: Not available

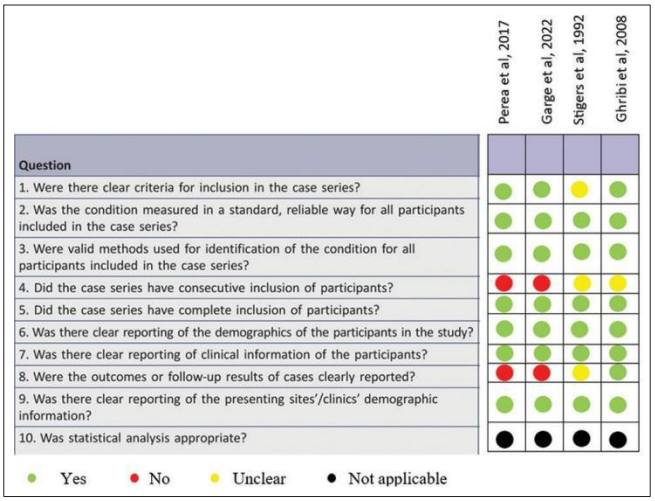


Figure 2: Methodological quality of case series studies – the Joanna Briggs Institute's critical appraisal tool

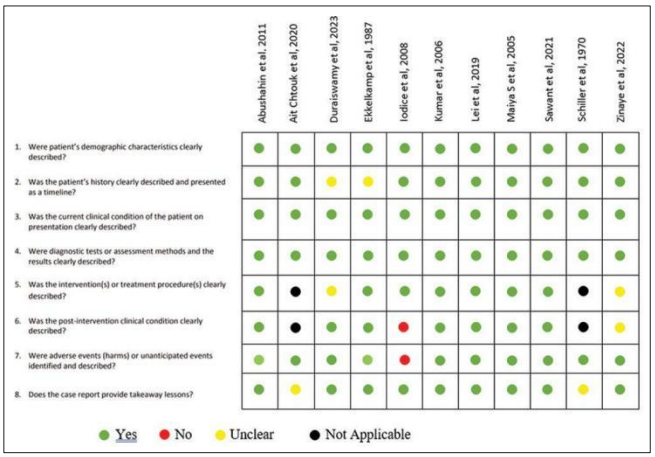


Figure 3: Methodological quality of case reports – Joanna Briggs Institute's critical appraisal tool

other diagnoses, such as pneumonia, acute bronchiolitis, asthma, congenital heart disease and early onset neonatal sepsis, were considered before arriving at the correct diagnosis. None of these reports had a mention about antenatal diagnosis of BCLE.

Lobar involvement pattern

Lobar involvement pattern was also evaluated, and the distribution of left upper lobe with right middle lobe (RML) was the most common and was seen in 17 cases (n = 17), i. e 80.9% cases. Lingula and RML together were seen in one patient (n = 1%–4.7%), RML with left lower lobe (LLL) and right lower lobe (RLL) in one (n = 1%–4.7%), RLL with LLL in one (n = 1%–4.7%) and RML with LLL in one child (n = 1 4.7%) [Table 2].

Management options employed

The studies were then evaluated based on the management option employed while managing these cases. Medical management was utilised in a 10-week-old infant presented who was managed by a conservative approach, as the child did not develop progressive symptoms and showed an early improvement.[6] By 1 year of follow-up, the child remained asymptomatic.

Surgical intervention

Surgical resection of the entire affected lobe(s) was done in 18 (85.71%) cases out of 21 studied with two cases

succumbing to the disease before intervention and one child who was managed conservatively. Lobectomy was performed via an open thoracotomy in all cases. In those undergoing simultaneous resection of both affected lobes, either a bilateral thoracotomy incision (n = 4) or a clamshell incision (n = 1) was used.

Lobectomy of both lobes was planned either simultaneously (n = 5, 27.77%) or sequentially (n = 13, 72.22%). In the latter group (n = 13), where sequential surgical lobectomy was performed, the mean duration between the first and second lobectomy was 67.6 days (4–180 days). In 6 cases (46.15%), only a unilateral lobectomy was performed with five of these patients (38.4%) not requiring a second subsequent lobectomy. In the one remaining child with unilateral lobectomy, the child expired before the second surgery could be performed due to a lack of parental consent for the subsequent procedure.

Outcomes and mortality

Four children out of the 21 reported cases expired due to medical complications – of these, two patients died before any medical/surgical intervention could be done. The diagnosis in these cases was made on autopsy.[8,9] One patient died a month after simultaneous surgery for BCLE[10] and one died within 20 days after unilateral surgery, before the second surgery could be done due to parental refusal for the second procedure[11] [Figure 4]. The odds of death after surgical intervention are three times in the simultaneous group when compared to the sequential group (Odds ratio: 3.0, 95% confidence interval: 0.15–59.89; Z statistic 0.719, significance level P = 0.47), although the P value is not statistically significant [Table 3].

Follow-up

The follow-up duration was not mentioned in the majority of the studies. The range in those who mentioned a follow-up period was 6 months to 5 years.

Discussion

Various management approaches were identified for the management of BCLE as per the literature available. Although one child was managed conservatively, the majority of patients underwent a surgical excision of the affected lobe and it remains the mainstay of treatment for BCLE. Lobectomy has been performed via an open approach in all cases. However, with advances in minimally invasive surgery and the widespread availability of appropriately sized instruments and reports of successful thoracoscopic lobectomy in neonates and infants, thoracoscopic lobectomy for BCLE also seems possible in the future.

No study exists comparing upfront simultaneous bilateral surgery and sequential surgery in BCLE patients. Sequential surgery was planned in 13 of the 18 (72.22%) BCLE cases. Interestingly, five children out of 13 (38.4%) did not seem to require a second surgery at all, i.e. only one lobe was removed.

Table 2: Lobar involvement in bilateral congenital lobar emphysema cases

Lobes	*n* (%)
LUL, RML	17 (80.9)
Lingula, RML	1 (4.7)
RML, LLL, RLL	1 (4.7)
RLL, LLL	1 (4.7)
RML, LLL	1 (4.7)

LUL: Left upper lobe, RML: Right middle lobe, RLL: Right lower lobe, LLL: Left lower lobe

Table 3: Outcomes of simultaneous versus sequential surgical intervention for bilateral congenital lobar emphysema patients

Type of surgery	Survival	Deaths	Total
Simultaneous	4	1	5
Sequential	12	1	13
Total	16	2	18

Significance level P=0.47, OR: 3.0, 95% CI: 0.15–59.89, Z statistic: 0.719. OR: Odds ratio, CI: Confidence interval

In the rest of the seven cases out of thirteen (53.8%), both lobes were successfully removed sequentially. This brings into question the role and the need for an upfront simultaneous bilateral surgery.

As far as simultaneous surgery in a neonate/infant requires careful patient selection, expert paediatric anaesthesia support and good post-operative intensive care. The report by Iodice *et al*.[12] has given the first description of the anaesthetic management of a patient undergoing surgery for bilateral CLE. The authors cited the concerns that the initiation of positive pressure ventilation would cause hyperinflation of the emphysematous lobes and compromise oxygenation, venous return and cardiac output. In their case, simultaneous BCLE excision was done via a clamshell incision, the only report that mentions this procedure. Bilateral open thoracotomy has the potential to cause ventilation problems in the intraoperative period but also having a bearing on the post-operative lung expansion and would demand a certain period of mechanical ventilation, adequate analgesia and even prolonged sedation, adding to the morbidity of the condition, thereby increasing the risk of complications.

On the other hand, sequential surgery was seen to be better tolerated, with the second surgery done in the majority of the cases only when the infant gets symptomatic for the contralateral side. The timing between both surgeries ranged from 4 days to 180 days (median 18.5 days). The rationale put forth for sequential surgery varied among the authors. Ghribi *et al*.,[7] felt that infants tolerate sequential surgery better; sequential surgery was done in their case. Duraiswamy *et al*.,[13] reported that their child had CLE diagnosed only on one side at first and when these children became symptomatic again, a contralateral side CLE was detected and tackled. Five other authors who opted for the sequential surgery route operated

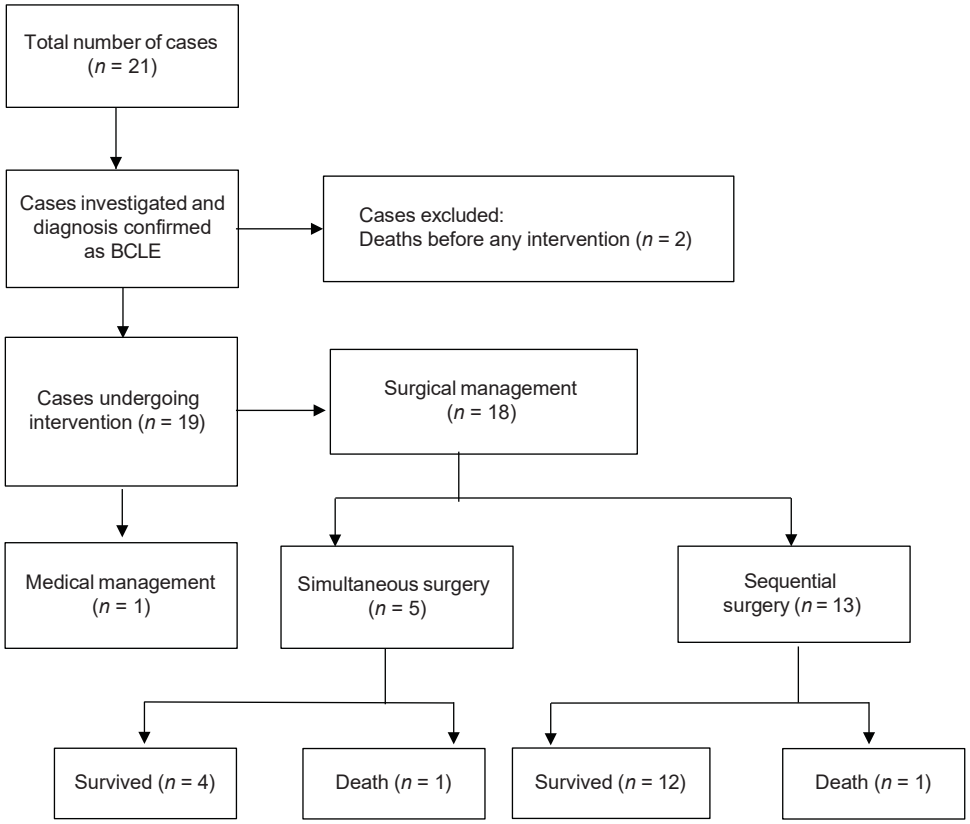


Figure 4: Summary of number of bilateral congenital lobar emphysema cases, their interventions and outcomes

on their BCLE when they turned symptomatic for the other side.[4,5,14,15]

The literature also points out that a planned sequential surgery did not preclude a possible decision to operate on the contralateral side during the same sitting. Sawant *et al*.,[16] had reported that their initial plan was to do a two-stage procedure, but their 4-month-old infant had persistent desaturation on the table up to 80%, for which they had to proceed with a contralateral lobectomy and saturation promptly improved.

Conservative management of BCLE has been reported and may be an option in carefully selected cases. Stigers *et al*.[6] described one patient who presented at 10 weeks of life, clinical and radiographic features were suggestive of CLE involving the RLL with the LLL. This case was managed by a conservative approach as the child didn't develop progressive symptoms and was asymptomatic on follow-up.

A total of 4 out of 21 reported cases have expired due to medical complications – of these, two patients died before any medical/surgical intervention could be done. The diagnosis in these cases was made on autopsy.[8,9] Kumar *et al*.[10] reported one death after simultaneous surgery performed on an infant who had a history of respiratory complaints since 1 month of life and was diagnosed with BCLE who expired 1 month after the surgical procedure. Lei *et al*.[11] reported one death after a unilateral surgery for BCLE on an infant who was diagnosed at the age of 77 days of life and operated on day 79. The child developed respiratory distress but the subsequent lobectomy was not performed due to parental refusal.

The management of BCLE remains challenging, with no consensus regarding the optimal approach. Surgeons have opted for various treatment options, including conservative management, simultaneous lobectomies or sequential surgical procedures. Surgical resection, particularly lobectomy, remains the mainstay of the management of these children. A sequential surgical management approach demonstrates promise in managing these difficult cases. Non-surgical management may be considered in less severe cases or as adjunctive therapy. A limitation of this review is that the summary has been synthesised from case reports and case series only, and this is primarily due to the extreme rarity of this condition. Further research, including prospective studies and comparative trials, is required to establish standardised treatment protocols and study the long-term outcomes of BCLE patients.

Conclusion

This systematic review highlights the various management strategies employed in the treatment of BCLE. Surgical resection remains the cornerstone of therapy, although the timing and extent of surgery vary among clinicians. We suggest sequential surgery be preferred for infants with given the current state of evidence, as the chance of not needing a subsequent surgery may be as high as nearly 40%. Conservative management may have a role in very select

cases. Future research should focus on the comparison of type of surgical approaches and the evaluation of the long-term outcomes of each of them.

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Conflicts of interest

There are no conflicts of interest.

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