

Lung ultrasound: A useful additional tool in clinician's hands to identify pulmonary atelectasis in children with neuromuscular disease

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Abstract

Introduction and Objectives: Patients with neuromuscular disease (NMD) are often exposed to ionizing radiations which could be reduced if a noninvasive and reliable diagnostic method is identified. The major aim of this study was to compare the use of chest X-ray (CXR) with lung ultrasound (LUS) in pediatric patients with NMD, to identify pulmonary atelectasis (PA).

Materials and Methods: A prospective study was conducted on children affected by NMD. In all patients who underwent CXR, a LUS was also performed and results compared for the assessment of PA.

Results: Forty children affected by NMD were enrolled. Spinal muscular atrophy type 1 was the most common NMD, followed by spinal muscular atrophy type 2 and congenital myopathies. More than half of the subjects presented PA, more prevalent in the lung left lower lobes. LUS agreed with CXR results (negative or positive for atelectasis) in 31 patients (77.5%). In two patients, an agreement evaluation was not possible due to non-conclusive LUS. Conversely, LUS disagreed with CXR in seven patients, four of which were positive for atelectasis. If only complete agreement was considered, the statistical analysis between CXR and LUS showed: LUS sensitivity of 57%, LUS specificity of 82%, positive predictive value 80%, negative predictive value 61%.

Conclusions: This study suggests that the use of LUS should be recommended to early identify PA and reduce frequent ionizing exposition of these fragile patients. Finally, our study also suggests that LUS can provide relevant information for clinicians and respiratory physiotherapists.

KEYWORDS

imaging, lung atelectasis, lung ultrasounds, neuromuscular disorders, spinal muscular atrophy

1 | INTRODUCTION

Neuromuscular disease (NMD) is a group of disorders that affect the motor neuron, neuromuscular junction and muscle fibers. A common

feature is the progressive muscle weakness that compromises the function of the respiratory system, including lowering upper airway tone, impairing cough, secretion clearance and weakening chest wall support.¹ Children with NMD are prone to respiratory complications,

such as upper airway obstruction, pulmonary aspiration, recurrent respiratory infections, hypoventilation and atelectasis.

Pulmonary atelectasis (PA) is a collapsed and not ventilated region of the lung parenchyma. Three are the main causes of atelectasis: airway obstruction, compression of parenchyma, and increased surface tension due to surfactant deficiency or dysfunction.² In cases of asthma and lower respiratory tract infections, the most common cause of PA is airways obstruction due to inflammatory processes. Conversely, in NMD, atelectasis is the consequence of overlapping conditions: obstruction of the bronchial lumen, compression of lung tissue and weakening of the respiratory muscles. Respiratory impairment is the leading cause of morbidity and mortality in children with NMD; therefore, the early recognition of a lung injury, such as PA, is essential. The fragile condition of all patients with NMD results in an excessive use of imaging exams, such as chest X-ray (CXR) and computed tomography (CT), involving a frequent exposure to ionizing radiation. In literature, lung ultrasound (LUS) has already been described as a reproducible, low cost, radiation-free and reliable diagnostic technique that avoids resorting to continuous radiological examinations. The 2012 International evidence-based recommendations for point-of-care introduced the role of LUS in neonatal and pediatric patients.³ Several authors emphasized that the specific anatomical features of children, such as shorter thoracic width, thinner chest wall, and small lung mass make LUS examination easier in children than in adults.⁴ LUS allows detection of pleural effusion, pneumothorax, and interstitial pulmonary edema. It has also been considered for lung consolidations⁵ where the lung parenchyma becomes a good transmitter of ultrasound producing an image called "hepatization of the lung" and sensitivity and specificity of ultrasound to diagnose lung consolidation exceed 90%. Air bronchogram is identified by ultrasound as an internal hyperechoic punctiform image.⁶ The consolidation and bronchograms LUS images are the main features of atelectasis.⁷

The use of LUS in diagnosing pediatric diseases, as neonatal respiratory distress syndrome⁸ or pneumonia,⁹ has already been described. However, to the best of our knowledge, no papers have been published on the use of LUS in children affected by NMD.

The recurrence of life-threatening pulmonary complications in this very fragile population, requiring continuous radiological exams, highlights the urgent need to identify alternative, noninvasive, reproducible, and radiation-free diagnostic tools. (Figures 1 and 2).

The study has two purposes: the first and the major, to compare the use of CXR with LUS in pediatric patients affected by NMD, with the aim of identifying the presence of PA. The second, to analyze LUS sensibility and specificity compared with CXR in the detection of PAs in the same patient population.

2 | MATERIALS AND METHODS

The study was performed according to the principles of the World Medical Association Declaration of Helsinki. The study was approved by our local Ethics Committee (protocol n. 412, Bambino Gesù Children's Hospital, Rome, Italy). Parents and/or guardians signed a written informed consent before LUS was performed.

A prospective study was conducted on children affected by NMD and admitted to the pediatric pulmonology and respiratory intermediate care unit of our hospital, for a follow-up assessment or an exacerbation. In all patients who had undergone CXR for previous evidence of PA, or had never been submitted to CXR in their lifetime or presented a worsening of their respiratory picture, LUS was performed within 6 hours following the admission. CXR evaluation was made by a radiologist experienced in pediatric lung disease, while LUS was performed by a radiologist or respiratory pediatrician with an expertise in this technique. Only the pediatrician was not blinded to the patient's history and clinical course but, as the radiologist, was blinded to CXR results. Both exams were performed at bedside since mobilization of most patients was complicated by their clinical conditions.

An ultrasound machine (SIEMENS mod Acuson P-300) equipped with linear (5-12 MHz) and convex probe (1-8 MHz) was used. LUS was performed in the supine position and in the lateral one for posterior lungs examination. Each hemithorax was divided into an upper and lower half to localize the lung injury. CXR and LUS were

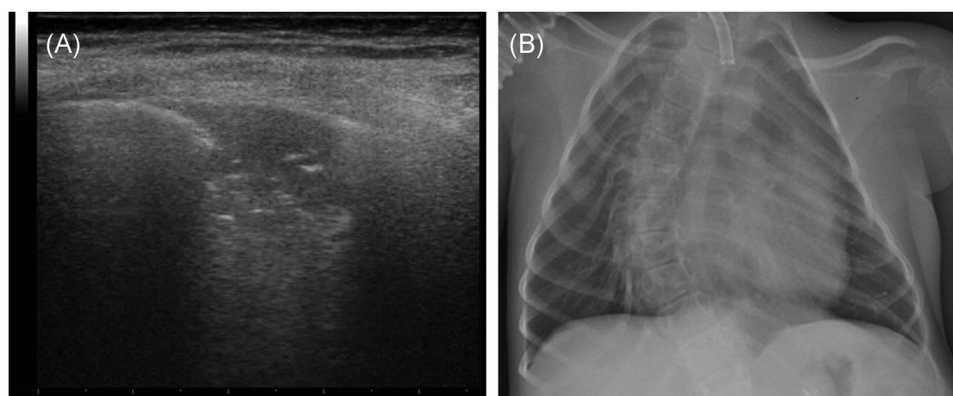


FIGURE 1 F,6 years, affected by spinal muscular atrophy type 2. A, lung ultrasound showed a large area of consolidation in the lower left lung. B, CXR confirmed atelectasis in the area of consolidation. CXR, chest X-ray

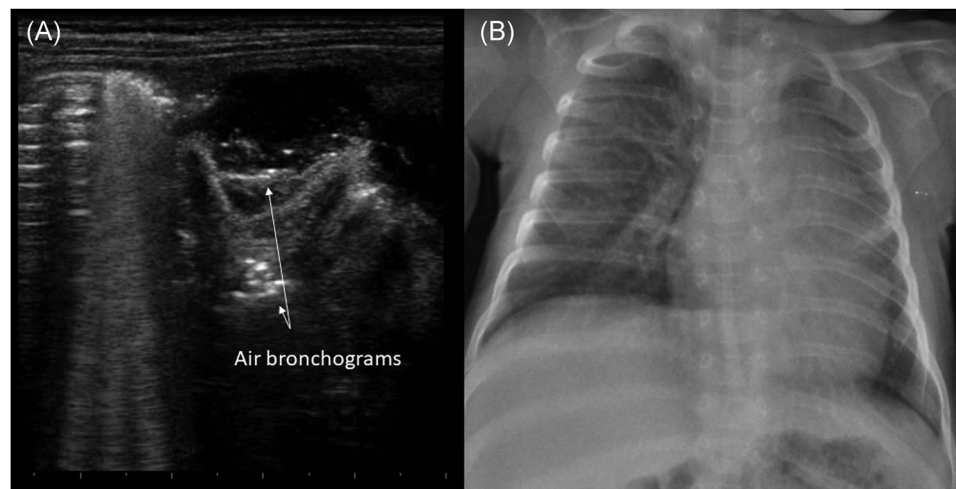


FIGURE 2 F,3 years, affected by merosin deficient CMD. A, lung ultrasound showed a large area of consolidation in the middle and lower left lung with air bronchograms. B, CXR confirmed atelectasis in the same lung area. CMD, congenital muscular dystrophy; CXR, chest X-ray

compared for recognition of PA and other lung injuries, such as pneumothorax and pleural effusion.

The statistical analysis was performed considering CXR as the gold standard for the diagnosis of atelectasis. The agreement between CXR and LUS was considered complete when both exams were negative or positive for atelectasis in the same lung areas. To analyze the agreement between the two tests, sensibility, specificity, and Cohen κ coefficient were calculated.

3 | RESULTS

A total of 40 children affected by NMD were enrolled. Population characteristics are described in Table 1. Spinal muscular atrophy type 1 was the most common NMD, present in 16 patients, followed by spinal muscular atrophy type 2 and congenital myopathies in seven cases. The median age was 7.5 years and 62% of patients suffered from comorbidities, the most frequent was severe scoliosis in 28% of them. More than half of the subjects presented PA, detected

by CXR in 22 patients (55%). Five patients presented more than one atelectasis, showing that NMD children are often affected by lung ventilation problems. LUS highlighted PA in 20 cases; in two patients LUS did not provide clear results because of severe scoliosis. As CXR, in five patients LUS showed a second atelectasis. PA were definitively more prevalent in the left lower lobes of the lung (59% of all cases of atelectasis were detected by CXR and LUS). Noteworthy is that LUS allowed to detect a modest pleural effusion (PE) in 15 patients, while CXR showed PE in only 10 subjects. Considering CXR as the gold standard, due to ethical reasons, CT was not performed. LUS agreed with CXR results (negative or positive for atelectasis) in 31 patients (77.5%). In two patients the agreement evaluation was not possible due to non-conclusive LUS, and in five patients the agreement was partial, because either the atelectasis position was different (three pts), or the number of atelectasis described was different (two pts). Conversely, LUS disagreed with CXR in seven patients, four of which were positive for atelectasis. In conclusion, the overall complete agreement between the two tests resulted to be in 65% of patients.

TABLE 1 Patient's characteristics according to different age groups

	<2 y	2-5 y	6-10 y	>10 y
Subject (n)	9	14	6	11
Sex (male), n (%)	7 (78%)	5 (35%)	0	7 (58%)
NMD (n)	SMA type 1: 7 SMA type 2: 1 Merosin deficient CMD: 1	SMA type 1: 9 SMA type 2: 3 CM: 2	SMA type 2: 2 DM1: 1 Myasthenia gravis: 1 CM: 2	SMA type 2: 1 SMA type 3: 1 DMD: 3 Ullrich myopathy: 1 Myasthenia gravis: 2 CM: 3

Abbreviations: CM, congenital myopathies; CMD, congenital muscular dystrophy; DMD, duchenne muscular dystrophy; DM1, myotonic dystrophy type 1; NMD, neuromuscular disease; SMA, spinal muscular atrophy.

If only the complete agreement was considered, the statistical analysis between CXR and LUS showed: LUS sensitivity of 57%, LUS specificity of 82%, positive predictive value 80%, negative predictive value 61%. The good agreement between CXR and LUS was confirmed by Cohen kappa coefficient, which resulted to be 0.63.

Moreover, LUS was performed by two different investigators (a radiologist and a respiratory pediatrician). When data were compared, agreement with CXR results did not show a significant difference between the two investigators: the radiologist reported 73% of agreement, while the pediatrician 63%.

4 | DISCUSSION

Our study shows that LUS could be considered a useful clinical tool to identify or rule out PA in pediatric patients affected by NMD. CXR was positive for atelectasis in 22 patients, while LUS in 20. This difference is explained by the presence of severe scoliosis in two patients. In fact, as scoliosis modifies the chest wall shape, it did not allow to correctly view the lung by LUS. This data supports what already reported by other authors, that LUS should not be only considered as a replacement technique of radiological exams, which offer valuable information, due to a wide panoramic view capability.¹⁰ In fact, the high prevalence of respiratory complications in children with NMD confirmed by our study (more than half of our population presented PA), explains the need for a full respiratory assessment and often radiological exams. However, this aspect highlights the importance of exploring LUS as noninvasive, reproducible and radiation-free technique. In these patients, LUS should be performed bedside as an extension of the physical examination, whereas radiological exams could be used to answer specific clinical questions when needed. LUS provides dynamic information which can be of relevant interest for an immediate diagnosis and overcome the need for many radiographies and CT scans,¹¹ the latter requiring child's difficult mobilization and involve a significant radiation exposure.

If only the complete agreement (when both exams identified the same number of atelectasis and in identical lung areas) was considered, the overall complete agreement between tests resulted in 65% of patients. Sensitivity and specificity showed LUS sensitivity of 57%, specificity of 82% with good agreement confirmed by Cohen kappa coefficient. To better understand our data and translate them into useful clinical practice, first of all we have to specify that considering CXR as the gold standard for ethical reasons, could have affected our results. In fact, patients with NMD often have severe scoliosis and an altered lung parenchyma, conditions that generate many artifacts in both (CXR and LUS). However, as authors experienced in the follow-up of patients with NMD, we believe that LUS can be considered a useful tool in clinician's hands to early detect the presence of atelectasis, thus suggesting, for instance, to increase respiratory physiotherapy and ventilation support. LUS, in expert hands, is an extra bedside tool to support the physical examination of

patients with NMD to get a deeper understanding of lung conditions and lead to a better respiratory management.

To our best knowledge, this is the first study exploring the use of LUS in children with NMD. Other studies showed the utility of LUS for neonatal PA,⁷ pediatric anesthesia-induced atelectasis¹² and pediatric cardiac surgery.¹⁰ LUS signs for atelectasis are alveolar consolidation and static air bronchogram. Alveolar consolidation has been described as a LUS sign for atelectasis but also of for pneumonia, while static air bronchogram is specific for atelectasis. In doubtful cases, clinical history and evaluation, other than blood tests, may be essential. The dynamic air bronchograms differentiate pneumonia from atelectasis with a specificity of 94% and a positive predictive value of 97% for diagnosis of pneumonia.¹³ For all kinds of alveolar consolidation, LUS sensitivity is 90% and specificity 98% compared with CT scan as the gold standard.¹⁴ In some patients with total or near-total opacification of the hemithorax, LUS has a high sensitivity in differentiating between consolidation from atelectasis and pleural effusion, whereas CXR is often unable to make this distinction.¹⁵ Our results and few studies in literature support the potential of LUS to detect or rule out PA. In addition, LUS can be used to evaluate the efficacy of therapies initiated for PA. The principles of managing respiratory complications in patients with NMD include secretion removal and treatment of possible aspiration, airway obstruction and respiratory insufficiency. Chest physiotherapy is used to mobilize secretions and treat ineffective airway clearance and aspiration through mechanical insufflation-exsufflation. CXR is not usually performed during a physiotherapist's intervention, while LUS can evaluate lung status in real time. LUS ability to distinguish between pleural effusion and lung collapse allows physicians and physiotherapists to identify recruitable lung areas. If an atelectasis is diagnosed, the implementation of lung recruitment techniques is highly recommended (manual or ventilator hyperinflation, PEP, body positioning, and expiratory-flow rate enhancement techniques) to promote secretion removal.

The resolution of atelectasis with increase in aeration can be proven by LUS with changes in the appearance of A lines and decrease of B lines. These changes allow to assess the effectiveness of the respiratory treatments.^{16,17} Therefore, unlike CXR, LUS could be used to make the initial diagnosis and guide the therapeutic approaches.

According to the diagnosis of pleural effusion, LUS showed PE in 15 patients, while CXR only in 10, thus confirming that LUS is a diagnostic tool more sensitive than CXR for the identification of pleural effusion in children with NMD, for both diagnosis and effusion quantification.¹⁸ Moreover, LUS can highlight pleural fluid characteristics, essential when a decision for drainage has to be made.

Finally, the agreement between LUS and CXR results did not show a significant difference when LUS is performed by a radiologist compared with the one carried out by a respiratory pediatrician. As already reported in literature, our results confirm that LUS can be a precious tool also in clinicians' hands.

The possible use of LUS for follow-up of atelectasis in patients with NMD was not investigated in this prospective study. However, we believe it should be considered for future research.

5 | LIMITATIONS

As previously said, the major limitation of our study is the decision of using CXR as the gold standard examination for the diagnosis of PA, instead of CT scan. However, both for ethical reasons and complying with the policy of reducing radiation/ionizing exposition, CT cannot be performed routinely, and its use must be limited to the most severe cases or when complications are suspected.

Another limitation is the relatively small number and heterogeneity of enrolled patients. Nevertheless, as NMD include a group of rare diseases with low population prevalence (1:3000), 40 children from a single center have to be considered a good representation of patients.

6 | CONCLUSION

This study suggests that the use of LUS should be recommended as a practical and additional bedside tool, as an extension of the physical examination, to early identify PA, get a deeper understanding of lung conditions and lead to a better respiratory management in young and fragile NMD children. All LUS limits, especially frequent artifacts, detected in this group of patients, often characterized by severe scoliosis and altered lung parenchyma, have to be fully understood and taken into account to avoid over diagnosis of atelectasis. The use of LUS has also the undiscussed positive effect of reducing, as much as possible, frequent ionizing exposition of these fragile patients. Finally, our study suggests that LUS can provide important information to both clinicians and respiratory physiotherapists.

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