Ultrasound of the pleura in children, WFUMB review paper

Joanna Jaworska¹, Natalia Buda², Ioana Mihaela Ciuca³, Yi Dong⁴, Cheng Fang⁵, Axel Feldkamp⁶, Jörg Jüngert⁷, Wojciech Kosiak⁸, Hans Joachim Mentzel⁹, Corina Pienar³, Jorge S. Rabat¹⁰, Vasileios Rafailidis⁵, Simone Schrading¹¹, Dagmar Schreiber-Dietrich¹², Christoph F Dietrich⁴,¹³

¹Institute of Mother and Child, Cystic Fibrosis Department, Warszawa, Poland, ²Internal Medicine, Connective Tissue Diseases and Geriatrics Department, Medical University of Gdansk, Poland, ³Department of Pediatrics, University of Medicine and Pharmacy “Victor Babes” Timisoara, Romania, ⁴Department of Ultrasound, Zhongshan Hospital, Fudan University, Shanghai, China, ⁵Department of Radiology, King’s College Hospital, London, United Kingdom, ⁶Pediatric Department, Sana Kliniken Duisburg GmbH, Germany, ⁷Department of Pediatrics and Adolescent Medicine, University Hospital Erlangen, Germany, ⁸Pediatric, Hematology & Oncology Department, Medical University of Gdansk, Poland, ⁹Section of Pediatric Radiology, Institute of Diagnostic and Interventional Radiology, University hospital Jena, Germany, ¹⁰Head Surgery Department Universidad de Oriente, Bolívar, Bolivar State, Venezuela, ¹¹Klinik für Radiologie und Nuklearmedizin, Luzerner Kantonsspital, Switzerland, ¹²Localinomed, Bern, Switzerland, ¹³Department Allgemeine Innere Medizin (DAIM), Kliniken Hirslanden Beau Site, Salem und Permanence, Bern, Switzerland

Abstract
Ultrasound (US) is an ideal diagnostic tool for paediatric patients owning to its high spatial and temporal resolution, real-time imaging, and lack of ionizing radiation and bedside availability. The lack of superficial adipose tissue and favorable acoustic windows in children makes US the first line of investigation for the evaluation of pleural and chest wall abnormalities. Lung parenchyma was previously thought to be inaccessible to ultrasound due to the presence of the air and bony thorax. The change in attitude and growing awareness of the diagnostic possibilities has led to lung ultrasound (LUS) being accepted as a valuable point of care method. In addition, the application of LUS has widened with improvements in technology such as higher resolution transducers, harmonic imaging and contrast-enhanced ultrasound.

In the current World Federation of Societies for Ultrasound in Medicine and Biology (WFUMB) paper series the topic will be introduced, the technical requirements explained and the use of ultrasound in the lung and pleura in pediatric patients are discussed.

Keywords: pleural effusion; atelectasis; pneumothorax; guidelines

Introduction
Ultrasound (US) is an ideal diagnostic tool for paediatric patients thanks to the optimal spatial and temporal resolution, real-time nature of imaging, lack of ionizing radiation and bedside availability. It is the first line of investigation for the evaluation of pleural and chest wall abnormalities owing to the lack of superficial adipose tissue and favorable acoustic windows in children. Lung parenchyma was previously thought to be inaccessible to US due to the presence of air and bony thorax. The change in attitude and growing awareness of the diagnostic possibilities resulted in lung ultrasound (LUS) being accepted as a valuable point of care method. In addition, the application of LUS has been broadened with improvements in technology: higher resolution transducers,
harmonic imaging and contrast-enhanced ultrasound. US also allow the diagnostic use of artifacts [1-6].

Chest ultrasound has been shown to be more accurate than chest radiography in identifying pulmonary oedema [7] and pneumothorax [8]. In a meta-analysis consisting of 1510 children, chest US showed significantly better sensitivity and similar specificity in detecting pneumonia in children compared to chest radiography [9]. It has been proposed as a reasonable alternative first-line investigation for diagnosing suspected community-acquired pneumonia [10].

The aim of this paper is to introduce into the technical requirements, the examination technique of the pleura, description of the normal pleura line, analysis of pleural effusion, diagnosis of pleuritis and differential diagnosis of benign and malignant pleural thickening in children.

1. Technical requirements

The technical requirements depend on the clinical question, the age, body size and the thickness of the subcutaneous tissue of the examined child. In older children and adolescents, whose anatomical structure is similar to that of adults, a convex transducer (3-7 MHz) is the first-choice probe (fig 1). In order to assess the details of the pleural line and the structure of the chest wall, a high-frequency linear transducer (7-20 MHz) should be used. In the case of younger children (especially neonates and toddlers), a linear transducer should be the first choice, possibly with the choice of trapezoid wide field-of-view imaging. Convex or micro convex transducers are used as supplementary probes.

For optimal chest and lung imaging, appropriate settings should be chosen, most importantly adjusting the depth of imaging, focus (at the pleural level) and turning off filters (eliminating or altering artifacts imaging) [1,2,11].

The colour and power Doppler options can be useful for assessing sub pleural consolidation, as well as lesions within the pleural cavity and chest wall [12]. Modern US devices also have a microvascular assessment module at their disposal, illustrating small vessels within solid lesions but contrast enhanced ultrasound is the only imaging method to display micro vessels and the enhancement pattern [13,14].

2. Pleura

The most common pathologies of the pleura or the pleural cavity are pleural effusion and pneumothorax. For the diagnosis of pleural disease, US has been a routine investigation for many years. This is because there is good accessibility of the pleura, especially the costal and diaphragmatic pleura. However, the paravertebral and mediastinal pleura are not adequately accessible with transthoracic US. US is a valuable imaging modality in children, particularly because of its lack of ionizing radiation. Furthermore, thoracic US is more sensitive than chest X-ray detecting small volumes of pleural effusion and is better than computed tomography in detecting thin pleural septations [15,16]. Knowledge of the typical presentations of pleural disease will help radiologists to better characterise pleural pathologies and to avoid diagnostic pitfalls.

2.1 Examination technique

Pleura and lung US should involve the entire accessible lung surface. The probe should be placed in the intercostal spaces in the body lines, parasternal, mid-clavicular, axillary, scapular, paravertebral, supraclavicular area and also over the abdominal organs (e.g. liver and spleen can be used as acoustic windows).

2.2 Normal sonographic appearance of the pleura

The normal pleura is 0.2-0.4 mm thick. Due to impedance differences the parietal pleura presents as a thin
hyperechogenic line. With high-resolution US probes, the parietal pleura presents as two separate layers, which correlates in fact with the thoracic fascia and the parietal pleura. The normal movement of the parietal pleura against the lung tissue during respiration is an important sign that aids diagnosis [17]. The visceral pleura is even thinner than the parietal pleura. In the situation where pathologic changes in the periphery of the lung results in no air spaces, the visceral pleura can be recognised as a thin hyperechoic line. The hypoechoic layer outside the parietal pleura correlates with fat tissue [18] (fig 2).

2.3 Pleural effusion

Pleural effusion is seen as an anechoic (sometimes echogenic) liquid formation with a sharp border to the pleura. Pleural effusion in children is most commonly diagnosed in the course of pleuropneumonia. Other underlying causes include: heart failure, kidney diseases, liver cirrhosis, pancreatitis, hypoproteinenaemia, fluid overload, trauma, congenital or acquired chylothorax and malignant effusion. Recent international pulmonary guidelines recommend the use of US of the pleura as the main diagnostic method for detection and characterisation of pleural effusion [19-21]. Larger effusions are easy to recognize, but very small pleural effusions in the costophrenic recess are sometimes difficult to distinguish from pleural thickening. However, with US, effusions with a volume of more than 5 ml can be detected with high sensitivity and specificity (average 100% and 99.7%) (fig 3).

This stands in contrast to the chest x-ray, where only effusions greater than 150 ml can be detected and x-ray sensitivity and specificity are moderate with an average of around 71% and 88% [21-23].

Pleural effusion can often be found posteriorly and at the lowest point dependent on the body position. Spleen and liver can be used as acoustic windows to examine the supradiaphragmatic aspects of the pleural cavities and the base of the lungs. If possible, the examination should also be done from the dorsal side of the patient, e.g. scanning the patient from the back when they are in a sitting position. The posterior axillary line is the preferred intercostal position for scanning over the medial and para-vertebral level. In doing so, the longitudinal section is shown intercostal. The entire thorax has to be examined as encapsulated effusions can occur anywhere within the pleural cavities. For the best resolution, a higher frequency linear probe should be used. In extensive pleural effusions a convex transducer might be helpful in order to obtain deeper penetration.

2.3.1 Determination of pleural effusion volume

Estimation of the volume of the pleural effusion is important for follow up and for the evaluation of therapy success. Many different estimation techniques are suggested which differ with regards to reliability and practicability. It is important to use the same tests for baseline and follow up to avoid any bias. Most reliable measurements are possible in a sitting patient with basal free expanding effusion but evidence-based studies are lacking in children. A good correlation has been described by multiplication of the effusion maximum height by a factor of 0.66. Another empiric formula for effusion calculation, which works well in clinical practice, multiplies the lateral effusion height at the chest wall in centimetres (cm) with the empiric factor of 90, which correlates with the volume of the pleural effusion in millilitres (ml)

Fig 2. Pleura anatomy. Thoracic wall with pleura visceralis (a, 1), pleural space (2) and pleura parietalis (3) in a 10-year-old boy with pneumonia. *Pleura visceralis appears thicker compared to the pleura in figure because of the total reflection of the normal underlying lung. Thoracic wall with pleura visceralis (b, 1), pleural space (2) and pleura parietalis (3) in a 4-year-old boy with septated pleural effusion in the pleural space. Especially the pleura parietalis is moderately thickened.

Fig 3. Small amount of pleural effusion (PE). The diaphragm (D) with three layers and the aerated lung (Lung) and the liver (Liver) are also shown. The acoustic phenomenon at the visceral pleura is seen as well.
Another practical technique multiplies the basal lung-diaphragm distance and the lateral effusion height with the factor 70 \((r=0.87)\) [24,25] (fig 4). Calculation of the effusion volume in a supine patient is significantly less accurate. An estimation formula gives the volume of the pleural effusion in ml by multiplication of the thickness of the pleural effusion in mm between the lung and the dorso-lateral chest wall in the posterior axillary line with the factor of 20 [26]. An easy estimation with good sensitivity (94-100%) but moderate specificity (76-67%) describes that a dorsobasal effusion thickness with left greater than 45 mm and right greater than 50 mm correlates with more than 800 ml volume of pleural effusion [27]. A large number of alternative methods are available. Regardless of which method is used, it is important to establish a uniform measurement method in the entire department, to ensure reliable and comparable measurement results through different sonographers.

### 2.3.2 Types of effusion

For adequate treatment, it is important to know the type of effusion. Transudates are complete anechoic. Exudates with high protein concentration and containing high numbers of cells or haemorrhagic effusions can be anechoic or may contain hyperechoic internal signals, which can float in a circle manner during breathing. Homogeneous hyperechoic effusions are especially seen in pleural empyema or haemothorax. Additional septations, pleural thickening or nodular pleural changes always suggest the possibility of an exudate. These are considered as “complicated” pleural effusions. These encapsulated or septated effusions are associated with a high risk of complications. Different forms of pleural effusion are shown in figures 5-8.

Haemothorax shows a variety of morphological changes depending on the stage [28]. Kozaci et al compu-
pared US and CT for identification of thoracic injuries. They found US highly specific and moderately sensitive in detecting thoracic injuries. While pneumothorax was identified with the highest sensitivity, the lowest sensitivity was observed for detecting haemothorax and subcutaneous emphysema [28]. The presence of subcutaneous emphysema is suggested by the presence of the E lines using the linear probe. E lines are vertical lines that reach the edge of the screen but do not arise from the pleural line [28].

Pleural empyema presents on ultrasound as an encapsulated not free expanding effusion with mostly homogeneous, internal signal and only as minimally or moderately hyperechoic. Pleural thickening occurs associated with inflammation of lung structures (including empyema and tuberculosis), but also with primary or secondary tumours. Conclusions regarding the aetiology based on the morphology of the pleural thickening cannot be drawn by US.

2.4.2 Pleuritis (diffuse pleural thickening)

The majority of patients with pleuritis show sonographic detectable changes (64-89%) [32]. Typical imaging findings are diffuse hypoechoic irregular pleural thickening, band-like fibrin manifestations are seen as hyperechoic thin linear changes within the pleural effusion based on fibrin strings, small sub pleural nodules (0.2-1.0 mm) or a lack of pleural gliding [32]. Only in 23% can increased vascularity can be observed on colour Doppler US. But with contrast enhanced US the hyperaemia of the pleuritis can be visualised in almost all cases [33,34]. However, due to the limited therapeutic consequences especially in children this technique has only a limited clinical relevance.

2.4.3 Benign and malignant pleural tumours

Pleural tumours are very rare in children. Most pleural tumours appear as a solid mass with broad adherence to the pleura on ultrasound. Often an adjacent pleural effusion is present.

Benign tumours in children include lipoma, neurinoma or chondroma. A benign neoplasm, which occurs typically in children and young adults, is the calcifying fibrous pseudotumour [35]. Previously, these tumours were termed as “childhood fibrous tumour with psamomma bodies”. A history of inflammation is a prerequisite for the diagnosis. On imaging, extensive solitary or multifocal masses with calcification are seen [36]. Another cause for pleural thickening in children are angiomas, which can be categorised easily as a vascularised mass with a chaotic blood flow on Doppler ultrasound. The majority of benign pleural tumours are moderately hyperechoic, circumscribed, show a thin capsule and contain calcification. They can replace or displace the surrounding lung tissue, but normally do not present with an infiltrating growth pattern. Small pleural effusions in the basal recesses or locally around the tumour are observed in benign pleural lesions and not only in malignant ones [37]. A reliable distinction of the different benign entities
is not possible with ultrasound. Primary malignant pleural lesions are even rarer in children than benign tumours (fig 9).

Typical imaging findings in malignant pleural tumours are effusion, pleural nodules, focal or diffuse pleural thickening and increased vascularisation on Doppler US. Cystic/necrotic changes and calcification are often seen after chemotherapy. Malignant lesions can be irregular, but many malignant pleural lesions are circumscribed, moderately hyperechoic without infiltration [38]. Accordingly, it is often impossible to distinguish them from benign pleural tumours, which underlines the need for minimally invasive biopsy for histological verification in solid pleural masses.

By far the most frequent malignant pleural lesions, but still very uncommon, are pleural metastasis and mesothelioma and other rare neoplasia [38]. Metastases may be found especially in patients with Wilms tumours and sarcomas. Both Hodgkin’s and non-Hodgkin’s lymphoma can involve the pleura. Pleuro-pulmonary blastoma is a very rare, malignant and highly aggressive tumour that originates from either the pleura or the lungs. It occurs mainly in children aged less than six years (90% between 0-2 years old). It has a poor prognosis with three different subtypes: cystic (type 1), combined cystic and solid (type 2) and solid (type 3). Pleuro-pulmonary blastomas are usually right-sided with contact to the pleura, usually without chest wall invasion or calcification. Blastomas have no characteristic findings on ultrasound and may present with a cystic lesion or a large region of consolidation without sonographic air bronchograms. It is impossible to differentiate pleuro-pulmonary blastomas from types 1 and 4 congenital pulmonary airway malformations (CPAMs) [39].

2.5 Fibrogenic changes of the pleura (Fibrothorax)
Fibrogenic changes, also called fibrothorax, of the pleura, in its early stage can be completely hypoechogenic and can imitate a pleural effusion. However, fibrogenic changes do not show movement during breathing because of adhesions between the pleural layers. Long lasting fibrogenic changes tend to be more hyperechoic and develop coarse heterogenic calcification [40,41].

2.6 Pneumothorax
Pneumothorax refers to the presence of air in the pleural cavity and exerts pressure to collapse the lung. The diagnosis of pneumothorax is important in neonatology. The incidence of pneumothorax in neonates is only 1–2%, however the rate may be as high as 30% in patients with underlying lung diseases or mechanical ventilation [42,43]. Traumatic pneumothorax is caused by blunt or penetrating trauma. Iatrogenic pneumothorax is a complication of diagnostic or therapeutic procedures, such as central line placement or mechanical ventilation, especially in premature neonates [44].

LUS is considered to be as accurate as the chest X-Ray for diagnosing pneumothorax [45]. In a multicenter study that included 42 infants, LUS had a sensitivity, specificity, positive predictive value and negative predictive value of 100% for the diagnosis of pneumothorax [46]. In comparison with lung ultrasound, sensitivity and specificity in diagnosing neonatal pneumothorax were 100% and 100% for LUS, 96% and 100% for CXR, and 87% and 96% for translumination [45]. These results prove the value of ultrasound in routine clinical practice when performed by experienced operators. Advantages in terms of reduced complexity, feasibility at the bedside, absence of exposure to ionizing radiation and detection of radio-occult pneumothorax make lung ultrasound the method of choice in several common clinical situations in children.

Free air in the pleural cavity tends to collect at the highest point, making a ventral examination necessary when the patient is in a supine position. A dorsal exami-
nation is usually unnecessary. A high-frequency linear transducer (10-17 MHz) is used. The thorax is scanned in the midclavicular, parasternal and anterior axillary lines from caudal to cranial in order to identify the extent of pneumothorax. Additionally, the thorax must be scanned to obtain axial or slightly oblique images to the presentation of the ribs in order to define the lateral expansion (lung point). In terms of the device settings, it is important to turn off the harmonic imaging, because the presentation of pneumothorax depends on the evaluation of artifacts. The signs of LUS suggestive of pneumothorax are: the absence of pulmonary sliding, the presence of pulmonary point and the presence of the “stratosphere sign” in mode M. The pleural line and the A lines are also present and the B lines are absent. M-mode might be helpful under certain circumstances to illustrate the stratosphere sign [47]. It has to be taken into account that some of the authors do not use M-mode. In case of a partial pneumothorax the transition zone between the moving lung and the pneumothorax is called “lung point” (fig 10). The lung point sign is particularly specific for pneumothorax. Still, although specific, it is not “sine qua non” sign for pneumothorax. If the patient has a severe pneumothorax, it is impossible to find the lung point. In such patients it is impossible to assess pneumothorax size with LUS alone. Furthermore, the semi-quantitative US assessment of the lung point sign reliably classifies pneumothorax size when compared with radiographic measurements, especially for small sized pneumothorax [49]. When two lung point signs are visualised, the “double lung point” sign, a loculated pneumothorax may be present [49]. In pneumothorax, the hypoechoic space between the pleural layers and the movement of the pleural layers during respiration are lost, and therefore, there are no B lines, since this is an artifact that originates from the pleural sliding. The appearance of B lines in one segment rules out pneumothorax at that specific site [44,50]. The diagnostic accuracy of LUS is as good as that of radiographs for the diagnosis of pneumothorax, and even greater in small size [8,44-47,50,51].

2.6.1. Pitfalls

Subcutaneous emphysema

There are pitfalls that simulate the sonographic diagnosis of a pneumothorax. If the thoracic emphysema is pronounced, the pleural line cannot be displayed. Due to the subcutaneous air there is an irregular total reflection of the ultrasonic waves. As a rule, however, the arrangement of these artifacts is very irregular, almost chaotic. However, pleural gliding cannot be represented.

Lung cyst

A parietal lung cyst or pneumatocele (pseudocyst) can simulate a pneumothorax. The air-containing cyst forms the boundary layer with the pleura, which in turn shows multiple A-lines. B-lines are missing and pleural gliding may also be missing. The lung pulse is theoretically present, but it may also be absent due to adhesion of the pleural sheets.

Conclusion

In the late 80ies and early 1990s, LUS has been introduced mainly to determine pleural effusion. LUS has been slowly extended to more general pediatric applications. Herewith, current applications in pediatric pleural pathology have been summarized to further distribute the knowledge of ultrasound in the world of pediatric patients.

Conflict of interest: none

References

8 Joanna Jaworska et al

Ultrasound of the pleura in children, WFUMB review paper