US evaluation of Poland syndrome

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Purpose

Poland's syndrome (anomaly) is a rare congenital condition (1:30.000 lived born [1]), firstly described by Alfred Poland in 1841[2]. Sporadic are much more frequent than familial forms [3-5]. Etiology is still unknown, the most accredited cause is a vascular subclavian damage during early embryonic life [6-8]. It includes unilateral agenesis of the pectoralis major muscle associated with others omolateral malformations of the thoracic wall, costal ribs, mammary region and brachysyndactyly [9]. Phenotypical expressivity is extremely variable, as it ranges from mild esthetical defect to complex hand or thoracic deformities with functional impairment (Fig. 1). In addition, the anomaly has been observed in association with other syndromes and malformations [10-18]. The phenotypical features of the Poland syndrome has not yet been described in large series of patients.

The aim of this paper is to describe the US scanning technique and the main ultrasound (US) findings in a consecutive series of n=187 patients with Poland syndrome.
Methods and Materials

In this paper, we prospectively examined the pectoralis region of n=187 consecutive patients [age, mean ±SD 11.2 ± 8.9; median, range 8.6 (1.4; 54.8); 71(38%) females and 116 (62%) males] affected by Poland syndrome with 12-5MHz and 17-5MHz US. Patients were consecutive and recruited in approximately three-years from all of our country by an italian association of parents whose sons are affected by Poland syndrome. Based on US findings of the pectoralis major muscle, we arbitrarily subdivided these patients in three classes:

*type-I:* patients with complete muscle absence

*type-II:* patients who had the only clavicular head of the muscle

*type-III:* patients in whom the lower part of the sternocostal head and the abdominal head were absent.

Each of these patients was then examined to assess the presence or absence of the pectoralis minor muscle.

During US examination, we performed spectral Doppler analysis on the subclavian, axillary and internal mammary arteries, and recorded any abnormality related to the rib cage, ipsilateral hand and heart position. Data were described as means, standard deviation (SD) and medians with a range for continuous variables, while absolute and relative frequencies were used for categorical variables. Normality of distribution of continuous variables was determined using QQ plots and the Kolmogorov-Smirnov test. Parameters of the study groups were compared using #2 or Fisher exact test for categorical variables. Comparisons between groups and sub-groups were performed by Kruskal-Wallis or the Mann-Whitney U test, as appropriate. A p-value less than 0.05 was considered statistically significant, and all p-values were based upon two tailed tests.

Statistical analysis was performed using SPSS for Windows (SPSS Inc, Chicago, Illinois USA).
Results

The US examination of the pectoralis major muscle was performed using a standardized technique based on four scanning planes:

#1. sagittal parasternal to demonstrate the muscle as a long strip lying over the ribs and the intercostal muscles (Fig. 4A,B);

#2. transverse over the insertion of the pectoralis major tendon into the humerus and the myotendinous junction of the long head of the biceps tendon located just posterior to it (Fig. 5A,B);

#3. oblique transverse at the deltopectoralis triangle switching color Doppler on to demonstrate the position of the cephalic vein in its short-axis as a landmark to separate the deltoid from the pectoralis (Fig. 6A,B);

#4. sagittal at the midclavicular line, placing the probe just caudal to the bone, to identify the subclavius muscle and a possible residual clavicular part of the muscle (Fig. 7).

Type-I patients with complete agenesis of the pectoralis major were 84/187, accounting for approximately 44.9% of cases. In these cases, sagittal parasternal planes (#1) did not reveal any muscle tissue superficial to the intercostal muscles and the ribs (Fig. 4C), the tendon was absent (#2) causing anterior subluxation of the myotendinous junction of the long head of the biceps (Fig. 5C,D) and there was no muscle tissue medial to the cephalic vein (#3) at the deltopectoralis space level.

Type-II patients had hypoplasia of the pectoralis major with absence of the sternocostal and abdominal heads and presence of the clavicular one. They were 83/187, accounting for approximately 44.4% of the total population. In these cases, the sternocostal and abdominal heads of the muscle were not visualized on sagittal parasternal planes (#1) (Fig. 8). The clavicular head was visible and could be distinguished from the more lateral deltoid muscle based on the position of the cephalic vein (#3). This vein was a critical landmark to avoid confusion between type-I and type-II patients. Sagittal midclavicular planes (#4) were also helpful to distinguish the clavicular head from the subclavius muscle, which is located more posteriorly and in close proximity with the clavicle (Fig. 7). In these patients, a smaller tendon of the pectoralis major was invariably detected (tendon thickness range Type-I = 0.8mm-1.2mm; contralateral side = 1.3mm-1.9mm) (Fig. 6C).

Type-III patients in whom the lower part of the sternocostal head and the abdominal head were absent were 20/187, accounting for approximately 10.7% of cases. Side comparison on #1 scanning planes demonstrated a shorter muscle with loss of the most caudal fibers. On #2, #3 and #4 scanning planes, no abnormality was observed.
Subgrouping the patients as above, no statistically significant (p=0.34) difference in gender was observed among the three classes. Basically, in most cases of our population, the anomaly was right-sided (right-sided = 62% vs. left-sided = 37.5% vs. 0.5% bilateral). This is in agreement with the literature which reports that the right side is more often involved. Then, subdividing patients depending on the right or left-sided disease, no significant difference was found concerning the distribution of the three classes in the two groups (p=0.79). Concerning the pectoralis minor muscle, it was observed in only a minority of cases (8/187 - 4.3%), but it was specifically found in the incomplete forms, with 3.6% prevalence in type-II disease and 25% prevalence in type-III disease (p=0.0001). In other terms, the lower the degree of hypoplasia of the pectoralis major the higher the prevalence of the pectoralis minor muscle. No significant side difference was observed in blood vessel size and arterial blood flow by comparing the Doppler spectra of right- and left-sided arteries at the thoracic outlet. As regard other anomalies there was no significant correlation between the three types of the muscle and rib abnormalities (p=0.94), unilateral nipple absence (p=0.64), sternal abnormalities (p=0.47), and destrocardia (p=0.43). US was able to depict costochondral anomalies in the ipsilateral rib cage. A spectrum of abnormalities ranging from hypoplasia/aplasia of single or multiple rib units was observed (Fig. 9). The incidence of upper limb abnormalities (p=0.03) and, more specifically, of hand abnormalities (p=0.02), including syndactyly and finger hypoplasia, was significantly higher in the group of complete agenesis of the muscle compared with the incomplete forms. Similarly, the complete agenesis of the pectoralis major was observed at a higher extent in kindreds with familiar transmission rather than in sporadic cases, although this figure did not reach statistical significance (p=0.07). Finally, the age of the diagnosis was earlier in type-I (1.1±2.4 years form birth) than in type-II (3.9±8.1 years) and type-III (8.2±11.6 years) muscle anomaly.
Fig. 1. A. Photograph of a 6 year-old child with Poland syndrome. Note the absence of the anterior wall of the axilla and the ipsilateral chest deformity due to the absence of the pectoralis major muscle and some costal defects. B. Photograph of left hand deformities in a 5 year-old child with complete agenesis of the pectoralis major.

Fig. 1: Fig.1

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Fig. 2. A. Schematic drawing and B gross cadaveric view of the anterior chest wall illustrate the anatomy of the pectoralis major muscle. The muscle consists of three heads - clavicular (1), stenocostal (2) and abdominal (3) - converging laterally to insert into the humeral shaft by means of a flat multilayered tendon. The pectoralis major muscle is separated from the deltoid (Del) by the deltopectoralis space (arrowhead) and the cephalic vein.

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Fig. 3. A. Schematic drawing and (B) gross cadaveric view of the anterior chest wall illustrate the anatomy of the pectoralis minor muscle (Pmin). This muscle is triangular in shape, arises from the III, IV, and V ribs, near their cartilages, and from the aponeuroses covering the intercostal muscles to insert into the coracoid (asterisk) with a flat tendon. In a more lateral position, the coracoid gives origin to the short head of the biceps tendon (SHBT) and the coracobrachialis (CoBr). Arrowheads indicate regional vessels.
Fig. 4. A,B, Extended field-of-view sagittal parasternal 12-5MHz US images over the pectoralis major muscle (in red). The muscle appears as a long strip lying over the ribs and the intercostal muscles (arrow). C. Extended field-of-view sagittal parasternal 12-5MHz US image in a patient with type-I anomaly shows complete absence of the muscle.
Fig. 5. A, Schematic drawing and (B) corresponding transverse 12-5MHz US image of the humeral insertion of the pectoralis major tendon (arrowheads). Note the relationship of this tendon with the myotendinous junction of the long (LHBT) and the short head (SHBT) of the biceps tendon, the coracobrachialis and the latissimus dorsi. C, Schematic drawing and (D) corresponding transverse 12-5MHz US image in case of absence of the pectoralis tendon (type-I) anomaly. Note the anterior subluxation of the long head of the biceps (LHBT) relative to the humeral shaft.
Fig. 6. A, Schematic drawing of the deltopectoralis triangle showing the position of the cephalic vein (arrowheads) as it courses alongside the pectoralis and the deltoid. B, Oblique transverse 12-5MHz US image shows the cross-sectional appearance of the cephalic vein (arrow). This vessel forms a separation plane between the pectoralis and the deltoid. C, Transverse 12-5MHz US image over the pectoralis tendon in type-II anomaly. The tendon (arrowheads) looks thinner than normal.
Fig. 7. A, B Sagittal 12-5MHz US image over the middle third of the clavicle in two patients with type-I (A) and type-II (B) anomaly. In A, the subclavius (SubCl) muscle is demonstrated as a tubular muscle slip parallel to the clavicle. No other muscle structure is visible superficial to it. In B, the clavicular head of the pectoralis major muscle (PMjCl) is found arising from the clavicle and lying over the subclavius. C, Schematic drawing of the subclavius muscle.
Fig. 8. A, B, Extended field-of-view sagittal parasternal 12-5MHz US image in a patient with type-II anomaly shows the normal clavicular head of the pectoralis major muscle (in red), whereas its sternocostal portion is absent.
Fig. 9. A, Sagittal 12-5MHz US image over the right anterior chest wall in a patient with type-I anomaly with (B) photographic and (C) CT correlation shows a hypoplastic cartilage (blue arrow) and the absence (orange arrow) of a rib.
Conclusion

US can complement clinical examination to assess the abnormalities of the pectoralis complex in patients with Poland syndrome. Partial forms with incomplete agenesis of the muscle are more common than clinically thought. Further experience is needed to determine if the presence of a rudimental clavicular belly and tendon of the pectoralis major may affect the clinical management.
References
