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Brachysyndactyly due to Poland Syndrome

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A 49-year-old woman, was referred for electroneuromyography examination for paresthesias, pain and thoracic outlet syndrome hypothesis. The motor and sensory nerve conduction and Electromyography were normal. At the physical examination we observed brachysyndactyly (Figure 1) and atrophy of the pectoralis major confirming the diagnosis of Poland Syndrome (PS).

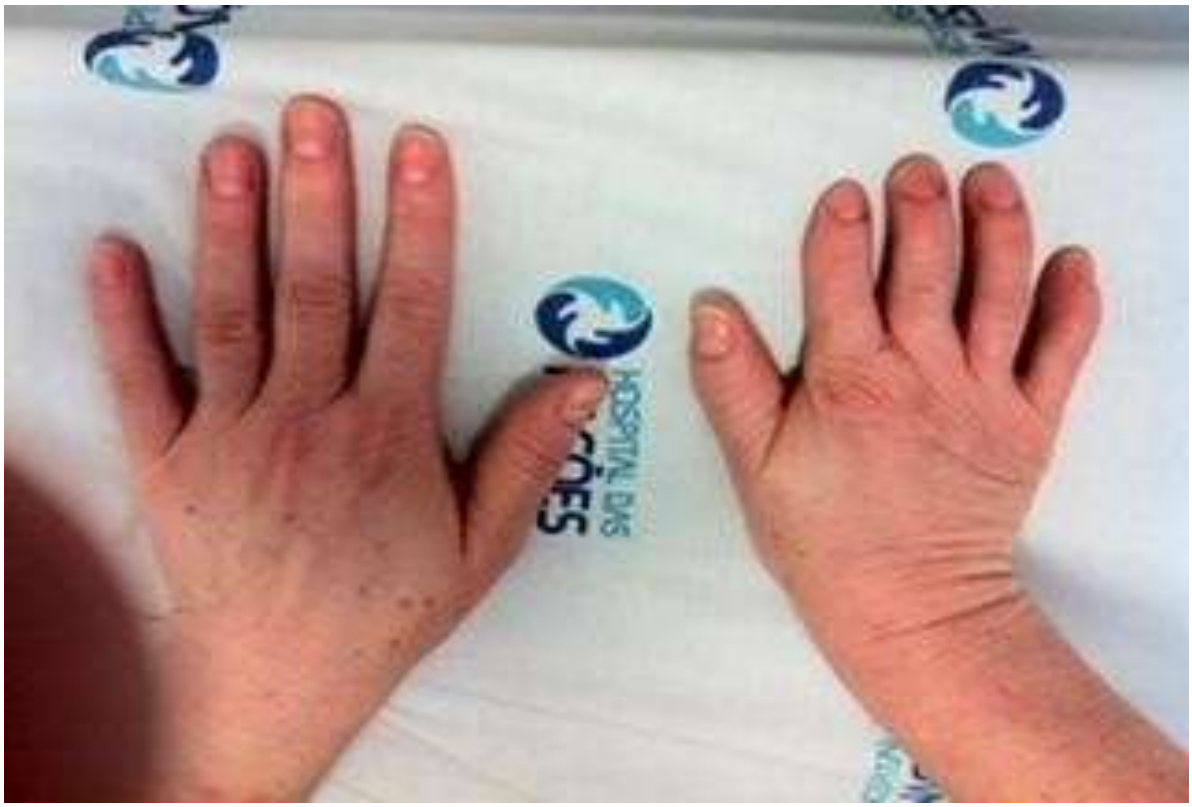

Figure 1. Brachydactyly of the right hand.

The first description is attributed to Alfred Poland, of Guy's Hospital, London, based on the autopsy of a 27-year-old male in which he described syndactyly of the hand with ipsilateral absence of the left sternocostal head of the pectoralis major. The condition was not named eponymously until 1962, when Dr. Clarkson, described a series of 3 cases of syndactyly with accompanying ipsilateral breast hypoplasia. [1, 2]

PS is a congenital syndrome characterized by deficiency of the pectoralis major muscle, presents a wide phenotype variability, including partial agenesis or deformity of the cartilage rib; hypoplasia or aplasia of the breast and nipple-areola complex, axillary fold, and subcutaneous tissue; sternal deformities. [3, 4]

The association of PS with the deformity of Sprengel, reported by 15.9%, may be one of the causes of referral for the performance of neurophysiological studies, since in the physical examination the carriers present elevation of the scapula. [3] Treatment of PS is determined by the severity of the defect and the resulting anatomical dysfunction. In conclusion, the early detection in clinical practice is important because it can be associated with muscle pain, joint pain and functional limitations, requiring an early multidisciplinary approach.

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