



Poland syndrome before Alfred Poland: the oldest medical description (Paris, France, 1803)

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Abstract

Here, we present a description of Poland syndrome from the second session of the Anatomical Society (Paris, France) on 11 December 1803 of congenital mammary absence and muscular atrophy on the right side. This case report predates the first official description of the disease published by Alfred Poland in *Guy's Hospital Reports* (London, 1841). Consequently, perhaps would it be necessary to do justice to its French discoverer, and to name from now on this nosological entity the “syndrome of Marandel”?

Keywords Congenital malformation · Retrospective diagnosis · Medical anthropology · Anatomical anomaly

Poland syndrome is a rare birth defect characterized by atrophy or absence of the chest muscle on one side of the body (mainly but not exclusively the pectoral). The prevalence of Poland's syndrome is 1–3 for 100,000 births [4]. It may be associated with ipsilateral syndactyly, shortened ribs, and anomalies of breast, nipple or axillary hair [3]. Pectoral muscle hypoplasia is not distinctive for Poland's syndrome alone, but is also present in syndromes with other associated anomalies with a recognized genetic cause. The prevailing theory on the etiology of this syndrome is an interruption of the blood supply to the subclavian arteries around the 46th day of embryonic development [7], but the pathogenesis is not clear and it is still being discussed: genetic, vascular disruption during embryogenesis, teratogenic effect or isolated defect in the paraxial mesenchyme during early limb development [2–9].

The first official report of this congenital anomaly was published by Alfred Poland (1822–1872) in the journal *Guy's Hospital Reports* (1841), after dissecting a 27-year-old convict (George Elt) who was said to have been unable to draw his hand across his chest due to deformity and ipsilateral hand anomalies [6].

No palaeopathological case has been described so far presenting the signs of Poland syndrome, but the congenital anomaly has been proposed as an explanation for the Amazon anomaly [1]. A potential case of Poland syndrome has also been suggested for a human depiction from Ancient Egypt (2400 BC), as a differential diagnosis of pectus excavatum [8].

Here, we present a case described in the second session of the *Société Anatomique de Paris* on 11th December 1803: “Congenital mammary absence on the right side. Citizen Marandel told the Society details of the dissection of the corpse of a woman who had a congenital mammary organ defect on her right side. A large portion of the sternohumeral [muscle] was missing from the same side as well as the entire costo-coracoid [muscle]. Besides, no alteration of the organs contained in the chest. All genital organs altered by the syndrome; each ovary presented a hydatid, the squirreous uterus adhered to the bladder, which was very contracted on itself, and presented internally small reddish tubercles, white within, easily cut. Citizen Ribes also told Citizen Dupuytren of a little girl with absolute absence of udders on one side. Citizen Laënnec says that authors

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without any medical background have cited cases of absence of the breasts” [5].

Such a clinical picture appears to correspond to the main traits of the disease: the atrophy of the muscles of the breast and of the associated glands, up to their cutaneous prolongations (areola, nipple). The association between this case and another survey by another contemporary practitioner makes it more sensible to think that some form of repetitive pattern might have been observed, unlike in Alfred Poland’s case, where only one sole autopsy case could be presented.

As a final reflection, it is stimulating to observe how Marandel’s description might really have predated Poland’s, in which the alternative eponyms of Marandel syndrome, or more conciliatorily, Marandel–Poland syndrome, may very well be introduced.

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Compliance with ethical standards

Conflict of interest No conflict of interest for all co-authors, relative to the subject of this article.

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