

## To The Editor:

I have read with interest the case report of "Congenital absence of multiple ribs" by Chen HJ.<sup>[1]</sup> For subjects with asymmetry of thoracic wall, Poland syndrome should be the first consideration. Until now, the etiology of Poland syndrome remains elusive. Interruption of the embryonic blood supply of the subclavian artery has been postulated, whilst the extent of involvement of the internal thoracic artery or brachial artery determines the severity of hand abnormalities.<sup>[2-4]</sup> Other causes of Poland syndrome may include disruption of lateral mesoderm,<sup>[5]</sup> single gene defects, trauma, viral infections or teratogenic effects of environmental xenobiotics, which have not yet been proven.<sup>[6-7]</sup> Since Dr. Chen did not mention whether the pectoralis major or minor muscles were absent, the ipsilateral subclavian artery was also not studied, we are not able to draw a definite conclusion from his presentation, in which the author argues that limb anomalies exclude Poland syndrome. Despite the first description of Poland syndrome in 1841 characterized by congenital deficiency of the pectoralis major or minor muscles, associated with symbrachydactyly, later references encompass a spectrum of Poland syndrome with variable associations of hypoplasia of breast, athelia and/or amnastia or hand anomalies.<sup>[8-10]</sup> Nonetheless, Poland syndrome should not be confused with anterior thoracic hypoplasia which refers to subjects with hypoplasia or depression of ribs but normal pectoral muscles. Subjects with pectoral hypoplasia without hand involvement are also thought to have "partial Poland's sequence".<sup>[11]</sup>

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## References

- 1 Chen HJ. Congenital absence of multiple ribs. *World J Pediatr* 2007;3:71-73.
- 2 Merlob P, Schonfeld A, Ovadia Y, Reesner SH. Real-time echo-Doppler Duplex Scanner in the evaluation of patients with Poland sequence. *Eur J Obstet Gynecol Reprod Biol* 1989;32:103-108.
- 3 Bavinck JNB, Weaver DD. Subclavian artery supply disruption sequence: hypothesis of a vascular etiology for Poland, Klippel-Feil, and Mobius anomalies. *Am J Med Genet* 1986;23:903-918.
- 4 Bouvet JP, Leveque D, Bernetieres F, Gros JJ. Vascular origin of Poland syndrome? *Eur J Pediatr* 1978;18:17-26.
- 5 Bamforth JS, Fabian C, Machin G, Honore L. Poland anomaly

with a limb body wall disruption defect: case report and review. *Am J Med Genet* 1992;43:780-784.

- 6 Freire-Maia N, Chautard EA, Opitz JM, Freire-Maia A, Quelce-Salgado A. The Poland syndrome—clinical and genealogical data, dermatoglyphic analysis, and incidence. *Human Hered* 1973;23:97-104.
- 7 David TJ. Nature and etiology of the Poland anomaly. *N Engl J Med* 1972;287:487-489.
- 8 Perez Aznar JM, Urbano J, Garcia Laborda E, Quevedo Moreno P, Ferrer Vergara L. Breast and pectoralis muscle hypoplasia. A mild degree of Poland's syndrome. *Acta Radiol* 1996;37:759-762.
- 9 Shamberger RC, Welch KJ, Upton J 3rd. Surgical treatment of thoracic deformity in Poland's syndrome. *J Pediatr Surg* 1989;24:760-765.
- 10 Spear SL, Pelletiere CV, Lee ES, Grotting JC. Anterior thoracic hypoplasia: a separate entity from Poland syndrome. *Plast Reconstr Surg* 2004;113:69-77.
- 11 Ireland DC, Takayama N, Flatt AE. Poland's syndrome. *J Bone Joint Surg* 1976;58:52-58.

## The Author Reply:

I am glad that Dr. Wong is interested in the case I reported recently. Dr. Wong considered that it is a case of Poland syndrome, which is worth discussing.

Poland syndrome is called "pectoralis muscle deficiency-syndactyly syndrome" that means pectoralis muscle deficiency and syndactyly, two concomitant physical signs at least, consist of the deformity, so Poland's syndrome is also named Poland's syndactyly. Cases of Poland's syndrome may be associated with hemangioma, leukemia, carcinoma of hypoplastic breast, and other conditions.<sup>[1]</sup> In recent reports,<sup>[2-5]</sup> the diagnosis of Poland's syndrome still includes limb anomalies like hypoplasia with syndactyly, brachydactyly and oligodactyly as described in *Smith's Recognizable Patterns of Human Malformation*<sup>[6]</sup>. Al-Qattan made a special classification of hand anomalies in Poland's syndrome,<sup>[7]</sup> but Foucras et al<sup>[8]</sup> analyzed 37 cases of Poland's syndrome and found malformations of hands in 12%. They considered that malformations of hands in Poland's syndrome are less frequent than the classical ones. Although there are different points of view for the diagnosis of Poland's syndrome, we have to follow the classical criteria for the diagnosis of the deformity before the new diagnostic criteria for Poland's syndrome are issued. This is why the case I reported has not been diagnosed as Poland's syndrome because of the absence of brachydactyly and syndactyly of her hands.

I appreciate Dr. Wong's opinion, which helps me to review in detail the relevant literature and better understand Poland's syndrome. I agree that the case may be diagnosed as "partial Poland's sequence" rather than "Poland's syndrome".

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### References:

- 1 Fokin AA, Robicsek F. Poland's syndrome revisited. *Ann Thorac Surg* 2002;74:2218-2225.
- 2 Kurt Y, Demirbas S, Uluutku AH, Akin ML, Celenk T. Poland's syndrome and gastric cancer: report of a case. *Eur J Cancer Prev* 2006;15:480-482.
- 3 Kulkarni ML, Sneharoop B, Vani HN, Nawaz S, Kannan B, Kulkarni PM. Eventration of the diaphragm and associations. *Indian J Pediatr* 2007;74:202-205.
- 4 Gerlinger I, Jarai T, Lujber L, Pytel J. Poland's syndrome and head-and-neck tumour: an unusual association causing a reconstruction dilemma. *Eur Arch Otorhinolaryngol* 2007; 264:553-556.
- 5 Riyaz N, Riyaz A. Poland syndrome (anomaly) with congenital hemangioma: a new association. *Indian J Dermatol Venereol Leprol* 2006;72:222-223.
- 6 Jones KL. Smith's recognizable patterns of human malformation, 5th ed. Philadelphia: WB Saunders, 1997: 116-302.
- 7 AL-Qattan MM. Classification of hand anomalies in Poland's syndrome. *Br J Plast Surg* 2001;54:132-136.
- 8 Foucras L, Grolleau JL, Chavoïn JP. Poland's syndrome and hand's malformations: about a clinic series of 37 patients. *Ann Chir Plast Esthet* 2005;50:138-145.