



Proteus Syndrome in the aetiology of carpal tunnel syndrome

We read with interest the article by N. Van Meir and L. De Smet on 'Carpal tunnel syndrome in children', in the October 2003 issue of 'Acta Orthopaedica Belgica' (4).

The authors should be congratulated for their excellent manuscript. A literature search had been made and the underlying aetiology of carpal tunnel syndrome (CTS) in children has been reviewed by the authors. They state that macrodactyly, 'a benign localised form of gigantism', is another unusual cause of CTS in children. We think that this could be part of other asymmetric overgrowth syndromes such as Proteus syndrome, rather than being a localised gigantism. In the aetiology of CTS, the authors could have mentioned Proteus syndrome, which is characterized by macrodactyly of hands and/or feet, skin naevi and subcutaneous-internal-peripheral nerve hamartomas (2, 3). Proteus syndrome is also of interest because of clinical overlap with the Klippel Trenaunay syndrome (1).

REFERENCES

1. **Biesecker L.** The challenges of Proteus syndrome : diagnosis and management. *Eur J Hum Genet* 2006 ; 14 : 1151-1157.

AUTHORS' REPLY

We have read the comments from Dr Saliha Senel with great interest. The correct diagnosis of overgrowth syndromes is challenging. There is an overlap between isolated macrodactyly, Proteus syndrome and Klippel Trenaunay. In the cases reported by us and those found in the literature, macrodactyly did not appear to be part of the full blown Proteus syndrome, mainly due to the absence of other characteristic features.

There is however a continuing discussion about the relationship between nerve compression and

2. **Choi ML, Wey PD, Borah GL.** Pediatric peripheral neuropathy in Proteus syndrome. *Ann Plast Surg* 1998 ; 40 : 528-32.
3. **Mc Cullagh M, Harper J, Pitt MC, Jones D, Spitz L.** Median nerve compression in Proteus syndrome. *Pediatr Surg Int* 1998 ; 13 : 449-450.
4. **Van Meir N, De Smet L.** Carpal tunnel syndrome in children. *Acta Orthop Belg* 2003 ; 69 : 387-395.

Saliha Senel, MD.

Department of Pediatrics, Dr Sami Ulus Children's Hospital, Ankara, Turkey.

Nurullah Okumus, MD.

Division of Neonatology, Dr Sami Ulus Children's Hospital, Ankara, Turkey.

Serdar Ceylaner, MD, Associate Professor.

Department of Genetics, Dr Zekai Tahir Burak Hospital, Ankara, Turkey.

Correspondence : Saliha Senel, Dr Sami Ulus children's Hospital, Telsizler, Ankara, Turkey.

E-mail: drsaliha007@yahoo.com.tr.

© 2007, Acta Orthopædica Belgica.

macroductyly, as some authors believe that the nerve overgrowth is the cause of the overgrowth of the soft tissues.

In contrast, we never saw hypertrophic nerves in Proteus syndrome.

Nathalie Van Meir, MD.

Luc De Smet, MD, PhD.

Hand Unit, Orthopaedic Department, University Hospital Pellenberg, Leuven, Belgium.