Journal of Pediatric Surgery (2012) 47, E1–E5

ELSEVIER

Journal of Pediatric Surgery

www.elsevier.com/locate/jpedsurg

Chest wall repair in Poland syndrome: complex single-stage surgery including Vertical Expandable Prosthetic Titanium Rib stabilization—a case report☆

Justus Lieber*, **Hans Joachim Kirschner**, **Jörg Fuchs**

Department of Pediatric Surgery and Pediatric Urology, University Children's Hospital, Tübingen, Germany

Received 4 June 2011; revised 4 August 2011; accepted 28 October 2011

Key words:
Poland syndrome;
Chest wall defect;
Autologous rib transposition;
Permacol patch;
VEPTR

Abstract Various surgical techniques have been described for repair of chest wall defects in Poland syndrome. We describe the case of a 16-year-old boy who underwent autologous rib transposition after sternal osteotomy. Chest wall stabilization was achieved using a combination of K-wires and Vertical Expandable Prosthetic Titanium Rib (Synthes GmbH, Freiburg, Germany). Reconstruction of the soft tissue defect was accomplished by combined latissimus dorsi muscle flap and Permacol patch (Covidien Deutschland GmbH, Neustadt, Germany). This approach might be considered an effective 1-stage treatment option of this condition in postpubescent boys.

Poland syndrome is a rare constellation of anomalies. Hypoplasia or absence of soft tissues, predominantly pectoralis muscles, constitutes the main clinical anomaly but may be combined with bony chest wall defects (eg, hypoplasia or absence of ribs) and upper extremity anomalies (eg, syndactyly or polydactyly). This syndrome was originally described by Alfred Poland in 1841. The incidence ranges from 1 in 7000 to 1 in 100 000. Males are affected more frequently with a 2:1 to 3:1 ratio. The right side of the body is involved in 60% to 75% of patients [1].

A variety of surgical techniques has been described for chest wall reconstruction depending on sex, age, and severity of the disease [1-7]. These approaches are frequently associated with disadvantages such as the use of exogenous implants, donor site comorbidities, and the need for multistage procedures. We describe a single-stage surgical approach that combines autologous ipsilateral rib transposition, Vertical Expandable Prosthetic Titanium Rib (VEPTR; Synthes GmbH, Freiburg, Germany) stabilization, and latissimus dorsi muscle flap over a Permacol patch (Covidien Deutschland GmbH, Neustadt, Germany).

1. Case report

A 16-year-old boy (Fig. 1A-B) with Poland syndrome without limb involvement presented with paradoxical movement of the right chest wall skin in the region of the defect, subjective perception of respiratory compromise during stress, and cosmetic as well as psychologic concerns. The patient's main objectives were the establishment of better thoracic

☆ Financial disclosure: none.

* Corresponding author. Department of Pediatric Surgery and Pediatric Urology, University Children's Hospital, D-72076 Tuebingen, Germany. Tel.: +49 7071 2986621; fax: +49 7071 294046.

E-mail addresses: justus.lieber@med.uni-tuebingen.de (J. Lieber), hans-joachim.kirschner@med.uni-tuebingen.de (H.J. Kirschner), joerg.fuchs@med.uni-tuebingen.de (J. Fuchs).

0022-3468/$ – see front matter © 2012 Elsevier Inc. All rights reserved.
doi:10.1016/j.jpedsurg.2011.10.077

Fig. 1 A 16-year-old boy with Poland syndrome. Preoperative status and postoperative results in the frontal (A, C) and lateral (B, D) view.

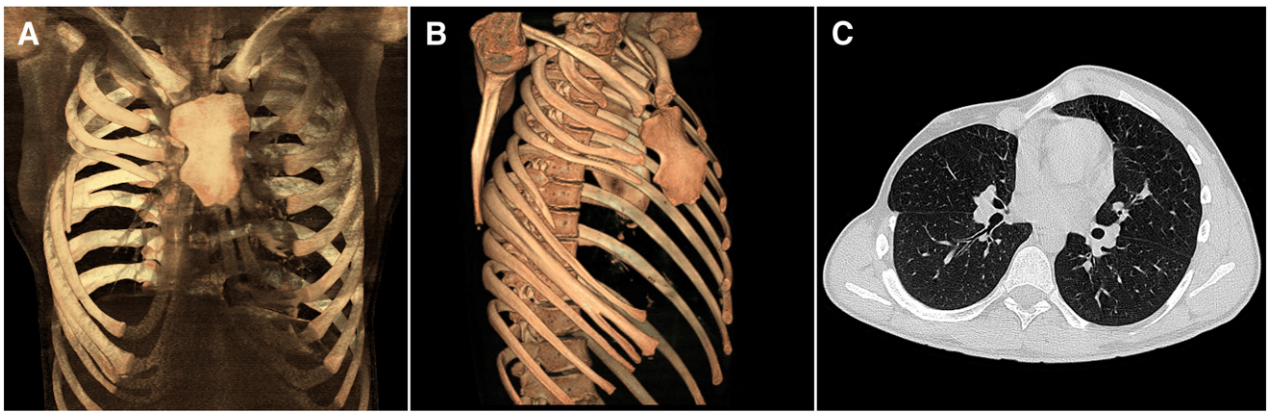

Fig. 2 Chest wall defect. Three-dimensional CT reconstruction including cartilaginous (A) and osseous structures only (B). Axial CT illustrates the right lung bulging toward the skin. The left-sided sternal buckle is also shown (C).