Expert's comment concerning Grand Rounds case entitled “Congenital kypho-scoliosis: a case of Thoracic Insufficiency Syndrome and the limitations of treatment” (by A.D. Chatterjee, K. Hassan and M.P. Grevitt)

Francisco J. Sanchez Perez-Grueso

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The authors describe a case of a 7-year-old girl with a severe spine deformity and repeated chest infections from birth [1]. On clinical exam, she presented a severe right-sided kypho-scoliosis and a stiff chest unable to expand. Oxygen saturation dropped to 85% when walking short distances.

X-rays showed on the coronal plane a right long curve including the thoracic and lumbar spine with the apex localized at the lower thoracic vertebra. The authors did not provide any information about the angular value of the deformity but it probably reached 80° Cobb angle.

The sagittal plane presented an angular kyphosis, over 90° Cobb angle. The apex seemed to be localized also at the lower thoracic vertebra, probably T10–T11.

3D CT reconstruction imaging showed multiple congenital anomalies affecting the spine and the rib cage, with

defects of formation and segmentation localized at the thoracic spine. The rib cage presented multiple bilateral fused ribs. No anomalies were found in the MRI.

Flexibility of this deformity was only assessed clinically under suspension that confirmed the rigidity of the kyphosis. The deformity was defined as Thoracic Insufficiency Syndrome because of the congenital multiple fused ribs and the patient's inability to maintain adequate levels of oxygen saturation while walking.

This patient was surgically treated with an anterior in situ fusion using two strut grafts from the mid thoracic spine to L2. The main aim was to prevent further progression of the deformity. The patient experienced an uneventful recovery retaking normal physical activity 1 year after surgery although the PFT did not improve.

Rationale for treatment and evidence-based literature debate

The authors considered that early surgery would be the best option for this 7-year-old girl in order to preserve lung and neurologic function by improving or preventing progression of the kyphosis.

After a complete review of the different techniques currently used in the treatment of early onset scoliosis, regardless the etiology, the authors came to the conclusion that standard procedures would not be suitable for this patient due to some specific factors affecting this case, such as presence of kyphosis, multiple anomalies, and potential risks of some techniques due to the severity of the deformity.

An anterior in situ fusion was considered to be the appropriate procedure to safely control the kyphosis. I fully agree with the authors regarding the main goal of treatment

F. J. Sanchez Perez-Grueso (✉)
Chair of Spine Surgery Unit,
Hospital La Paz, P^o de la Castellana 261,
28046 Madrid, Spain
e-mail: perezgrueso@gmail.com

but I would like to make some comments on the way they approached the case.

X-rays and 3D CT scan

The images provided in the manuscript show the lumbar spine with normal anatomy and the thoracic spine with multiple anomalies besides the congenital fusion involving most of the ribs in both sides. There are two fully segmented T10/T11 right hemivertebra, localized at the apex of the scoliosis and the kyphosis. The abnormalities of the posterior arch will determine the ultimate stability of the spine at this level. A careful view of the lower thoracic posterior elements shows hypoplasia of T10, T11 lamina. These two malformed vertebrae represent the junction zone between the flexible lumbar spine and the rigid long lever arm formed by the upper thoracic spine and the bilaterally fused ribs. This zone becomes very unstable and the spine collapses in angular kyphosis and in scoliosis [2, 3]. In summary, the deformity involves the whole spine, the upper part is congenitally malformed and rigid, and the lower part is normal and flexible, leading to a very unstable junction zone.

Treatment options

VEPTR technique [4]

This technique was originally designed for thoracic deformities with absent or fused ribs and for some congenital or syndromic conditions (Jarcho Levin and Jeune's syndrome). After a decade of experience its benefits in the treatment of sagittal deformities have not been proven. I agree with the authors that this technique is not the right procedure for the case despite the presence of multiple fused ribs.

Growing rods [4]

They were originally designed to control progressive early onset non-congenital scoliosis while allowing the spine to grow. They have proven to be effective in preventing curve progression although with a high rate of complications especially when kyphosis was present [5]. The benefit of growing rods in congenital deformities is limited due to the anomalous growth pattern existing in the congenital spine. There is no indication for this technique in the case.

Vertebral column resection (VCR) [6]

Vertebral resection allows substantial correction while keeping the length of fusion as short as possible. The best indication for VCR in children is a rigid and angular deformity. It is indicated when other surgical techniques

are not sufficient to obtain an acceptable correction. This technique is considered to be associated with a high risk of neurological complications. Increased experience in this salvage procedure and modern techniques of intraoperative spinal cord monitoring have significantly reduced the risk of neurological injury [7]. Although authors discarded this technique because of the unacceptable risk of spinal cord damage and the presence of too many vertebral anomalies for resection, I believe that the resection of T10 and T11 hemivertebra would be the right procedure to correct the deformity.

Selected technique

An anterior spinal fusion was done from the mid thoracic vertebra to L2 with a double femoral allograft held to the spine with two interference screws. The authors do not explain why they did not perform a posterior fusion to enhance the fusion mass. The posterior element defects found in the CT scan might be even due to image artifacts. The presence of some lamina hypoplasia would not contraindicate a posterior instrumented fusion including the same levels as the anterior fusion. It would protect the anterior struts grafts.

Outcome

It should emphasize the rapid postoperative recovery and the significant clinical improvement experienced by the child at 1-year follow-up despite the fact that the PFT did not change much. The authors expect PFT to return to normal values in a period of time of 2–3 years. I fear that this improvement will never happen given the type of the deformity and the age of the patient.

A close look at 1-year follow-up X-rays discovers that the deformity has worsened in both planes and probably will continue in the future unless surgical reconstruction is planned.

Personal preference

I would put this patient under halo traction, given the instability at the apex of the deformity, followed by a posterior VCR at T10–T11 levels and instrumented fusion.

References

1. Chatterjee AD, Hassan K, Grevitt MP (2011) Congenital kyphoscoliosis: a case of thoracic insufficiency syndrome and the limitations of treatment. *Eur Spine J* (submitted for publication)

2. Dubousset J (1994) Three-dimensional analysis of the scoliotic deformity. In: Weinstein SL (ed) *The pediatric spine: principles and practice*. Raven Press, New York, pp 479–496
3. Kawakami N, Tsuji T, Imagama S, Lenke LG, Puno RM, Kuklo TR, Spinal Deformity Study Group (2009) Classification of congenital scoliosis and kyphosis: a new approach to the three-dimensional classification for progressive vertebral anomalies requiring operative treatment. *Spine* 34(17):1756–1765
4. Yaczici M, Emanns J (2009) Fusionless instrumentation systems for congenital scoliosis: expandable spinal rods and vertical expandable prosthetic titanium rib in the management of congenital spine deformities in the growing child. *Spine* 34(17):1800–1807
5. Bess S, Akbarnia BA et al (2010) Complications of growing-rod treatment for early-onset scoliosis: analysis of one hundred and forty patients. *J Bone Joint Surg Am* 92(15):2533–2543
6. Lenke LG, O’Leary PT, Bridwell KH, Sides BA, Koester LA, Blanke KM (2009) Posterior vertebral column resection for severe pediatric deformity: minimum two-year follow-up of thirty-five consecutive patients. *Spine* 34(20):2213–2221
7. Hamzaoglu A, Alanay A, Ozturk C, Sarier M, Karadereler S, Ganiyusufoglu K (2011) Posterior vertebral column resection in severe spinal deformities: a total of 102 cases. *Spine* 36(5):E340–E344