Poland syndrome: A contraindication to the use of the internal thoracic artery in coronary artery bypass grafting?

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Poland syndrome (PS) is a rare congenital disorder that is present in 1:30,000 people. It is primarily characterized by hypoplasia of the pectoralis muscles, with associated ipsilateral syndactyly of the upper extremity. Although the true cause of PS remains unknown, the prevailing theory attributes these sequelae to hypoplasia of the subclavian artery or its branches as the result of an in utero vascular accident.^{1,2} Contemplating how to perform coronary artery bypass grafting (CABG) in a patient with left-sided PS is therefore of particular interest, because hypoplasia of the subclavian artery would theoretically render the left internal thoracic artery (LITA) inadequate for use. Consistent with yet exacerbating this clinical conundrum, no cases of CABG in patients with PS have previously been reported.

We recently encountered a patient with undiagnosed left-sided PS with significant coronary artery disease involving the left anterior descending coronary artery. Our preoperative workup included diagnostic imaging of the heart and great vessels to assess the adequacy of the LITA as a bypass conduit. Surprisingly, our evaluation demonstrated normal left subclavian artery and LITA anatomy, thus calling into question the currently accepted theory for this unusual syndrome.

Clinical Summary

A 70-year-old man was seen with anginal symptoms that had worsened during the previous 2 weeks. Physical examination revealed syndactyly of the left hand and hypoplasia of the left breast (Figure 1). Cardiac catheterization demonstrated three-vessel coronary artery disease, including a severely occluded left anterior descending coronary artery. After surgical consultation, the presence of the aforementioned phenotype was noted, and select subclavian artery catheterization was therefore performed. Interestingly, a widely patent subclavian artery and LITA were seen. Because of the possibility of chest wall deformity that might alter our sternotomy and closure, a computed tomographic angiographic study was also performed (Figure 2). This scan confirmed the absence of pectoralis musculature and demonstrated a normal bony

Figure 1. Syndactyly of left hand in PS.

thoracic cage. Moreover, normal caliber subclavian artery and LITA were confirmed. Intraoperative assessment of the LITA also revealed a normal artery with adequate flow by visual inspection. Four-vessel CABG was therefore performed, which included a LITA conduit to the left anterior descending coronary artery. The remainder of the operative and postoperative course was unremarkable.

Discussion

PS is a rare congenital anomaly first described by Froriep in 1839 and Poland in 1841. The clinical manifestations are extremely variable but are typically characterized by hypoplasia of the right breast, nipple, and pectoralis major muscle, combined with ipsilateral anomalies of the upper extremity, including brachysyndactyly and deformities of ribs 2 through 5. Familial PS is rare; spontaneous forms of PS are more common. Although numerous reconstructive procedures have previously been described in patients with PS, CABG has not previously been reported.

The currently accepted theory for the cause of PS centers on the end of the sixth week of development, when the upper limb bud adjacent to the chest wall is still in an early stage of development. It has been postulated that an interruption of blood flow during this critical time might subsequently lead to hypoplasia of the subcla-

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Figure 2. Computed tomographic angiogram with 3-dimensional reconstruction illustrating normal subclavian vasculature.

vian artery and its branches. Consequently, the extent and severity of the resulting phenotypic anomaly is thought to be causally related to the site and magnitude of the flow impairment within the associated vasculature.

This theory, although plausible, is supported by little evidence in the literature¹⁻³ and is clearly brought into question when

considering the normal vascular anatomy documented in our patient. If the proposed etiology is to remain accepted, it must be postulated that any in utero flow anomalies in this case were subsequently resolved before birth and therefore occurred only transiently during some critical period of organogenesis. Alternatively, a more tenable theory in light of our findings proposes that disruption of the lateral plate mesoderm (the embryonic origin of the anatomy affected) occurs, thus affecting the chest wall and upper extremity development between 16 and 28 days after fertilization.⁴

CABG performed with the LITA provides a known survival benefit to patients undergoing coronary artery revascularization.⁵ Use of this graft in patients with left-sided PS therefore would clearly be desirable, but there is a theoretic concern that subclavian hypoplasia might exist. To our knowledge, this brief communication represents the first and only report documenting normal vasculature before successful CABG with the LITA in a patient with left-sided PS. Although our findings give some cause for rethinking the etiology of this curious disease, individual demonstration of similar patency would be prudent before any proposed coronary revascularization in patients similarly affected.

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