Definition of the Inclusion Criteria of Poland's Syndrome

To the Editor:

I read with interest the invited commentary by Rodgers [1]. There are many thoracic anomalies characterized by the absence or hypoplasia of muscles and bones. In addition, some of these anomalies are seen together. The most noticeable example is Sprengel deformity, which was often accompanied with Poland syndrome. Many new variants of the thoracic anomalies such as Poland syndrome have been reported. Rodgers stated that all these disorders could be included under the rubric of “congenital thoracic musculoskeletal disorders” [1]. I completely agree with Rodgers’s views on the main title, which will cover all of these kinds of the anomalies.

With the exception of Poland syndrome, other thoracic anomalies have been defined precisely. Classically, Poland syndrome included the absence of pectoral muscles and ipsilateral hand anomalies. Over time, the other components and new variants of the syndrome have been reported. The main accepted component of the syndrome is partial or complete absence of great pectoral muscle. Agenesis of the costal cartilages and absence of the anterior parts of the ribs, abnormalities of the breast, lack of the subcutaneous tissue, and pectoral or axillary alopecia were reported as additional components of the Poland syndrome. However, there is a tendency to diagnose Poland syndrome in all patients with only isolated pectoral muscle agenesis or some thoracic deformities without the absence of pectoral muscle. In most reported case series, the patients with at least one or more defined components in addition to absence of great pectoral muscle have been considered to have Poland syndrome [2].

In the light of this information, the inclusion criteria of Poland syndrome should be restricted. Absence of great pectoral muscles might be defined as the major criterion of the syndrome, and the other accepted components might be defined as the minor criteria of the syndrome. Patients with a major criterion and at least two minor criteria should be considered as having Poland syndrome. These exact criteria will be helpful to solve the confusion in the diagnosis of the Poland syndrome.

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References

Reply
To the Editor:

I enjoyed reading Dr Yiyit’s thoughts [1] on my invited commentary [2], focused on his manuscript describing 8 patients with bilateral chest anomalies [3].

The difficulty in discussing Poland’s syndrome and many of the other thoracic musculoskeletal syndromes is our lack of understanding of the mechanisms of formation of these disorders. More and more medical geneticists are reserving the term syndrome for those anomalies for which we know the cause, usually genetic. In the absence of this understanding, they would prefer usage of terms such as association or complex.

Although clinicians have difficulty in giving up the appellation syndrome, this particular constellation of anomalies would be best described as Poland’s association. Most authors would require, at least, the presence of hypoplastic or absent pectoralis major and minor muscles and at least two of the various other anomalies that have been described for a patient to be considered as having Poland’s association. Accepting that classification, all 8 of the patients reported by Dr Yiyit and colleagues could be considered to have Poland’s association.

Let us bury the term “Poland’s syndrome,” and refer to it as “Poland’s association,” at least until we better understand the specific etiology of this disorder.

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The Left Thoracotomy Approach for Esophagectomy

To the Editor:

I appreciated the timely reminder of the utility of the left thoracotomy approach for performance of esophagectomy for selected patients as presented in the article by Ma and colleagues [1]. As the authors suggest, this versatile approach is probably underused. There are no components of an oncologically or functionally appropriate esophagectomy that cannot be accomplished. An uncompromised lymph node dissection of the abdomen is readily performed using this access. As there are no esophageal arteries arising from the aortic arch, the esophagus is easily mobilized to the top of the chest, so that a high thoracic (avoiding the free reflux associated with a low anastomosis) or a cervical esophagogastronomy can be constructed.

However, I want to urge an alternative to the authors’ recommendation of performing a “radial incision” of the diaphragm. Although this maneuver will provide access to the upper abdomen, as pointed out to me by my mentors Ron Belsey and David Skinner, there is a cost. This radial incision transects the branches of the phrenic nerve emanating from its terminus at the base of the pericardium and fanning out laterally. The result is an at least partially denervated diaphragm lateral to the incision and the dysfunctional sequelae thereof. Accordingly, I always heeded their advice to gain access to the abdomen through a peripheral incision of the diaphragm, parallel to and approximately 2 cm from its attachment to the chest wall. This