Frozen elephant trunk for aortic arch dissection in patients with Marfan syndrome: Are we there yet?

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Chen and colleagues from Beijing Anzhen Hospital investigated the remodeling of the distal aorta in patients with Marfan syndrome with type A aortic dissection (TAAD) treated with total arch replacement and frozen elephant trunk (FET). Their experience included 172 patients with an average follow-up of 6.2 years. Operative mortality was 8.1%. At hospital discharge, obliteration of the false lumen at the proximal end of the FET occurred in 98.7% of patients and it occurred at the distal end of the FET in 86.1% of patients. False lumen obliteration rate was lower at the level of the unstented thoracic aorta (39%) and the level of the renal arteries (21%). Over time, the true lumen of the distal aorta continued to increase, whereas the false lumen size remained relatively stable. Freedom from distal aortic dilation at 10 years was 57.6%, which was higher in patients with smaller preoperative aorta size and obliterated distal false lumen postoperatively. At the latest follow-up, complete aortic remodeling defined as obliteration of the false lumen and normal aorta size was achieved in 56.4% of patients at the level of the FET and 28.8% of patients at the mid-descending thoracic aorta.

With this large series with mid- to long-term follow-up data on the distal aorta, reasonable evidence is accumulating in support of the role of FET in the positive remodeling of stented aortas, specifically in patients with Marfan syndrome. The study also supports the ongoing notion that endovascular repairs can be performed safely in appropriately selected patients with connective tissue disorders. Although it appears that the false lumen is stabilized in unstented and distal thoracoabdominal aortas, complete remodeling is not achieved, which concurs with our current understanding of the natural history of the residual dissected distal aorta in patients without Marfan syndrome.

Despite the provocative message of the article, the results should be interpreted with some caution. First, it must be noted that this is a heterogeneous series that included patients with both acute (54.7%) and chronic (45.3%) TAAD from a single surgery center. It is remarkable that they had such a large number of patients with Marfan syndrome with chronic TAAD. It is likely that patients with Marfan syndrome who survive the acute event and go into the chronic phase have a different aortic pathology and natural history than those who do not. In fact, patients with acute TAAD had more rapid growth of the distal aorta than did the chronic group. Second, it is notable that complete aortic remodeling was not demonstrated to be associated with higher late survival or freedom from reoperation. The question of whether radiographic success translates into improved clinical outcome remains to be answered. Studies with longer follow-up and appropriate statistical power and methodology will be required to answer this important question.

Despite these limitations, this study adds evidence supporting the durable role of FET in the treatment of TAAD in patients with Marfan syndrome by providing positive aortic remodeling in the stented portion in the majority of treated patients and stabilizing the distal false lumen. We believe that FET should hold a role in a strategy for staged and hybrid total aortic replacement in patients with Marfan syndrome. It can potentially reduce the extent of subsequent thoracoabdominal aortic replacement with implications on stroke and paraplegia rates, but this has yet to be shown.
What is certain is that cardiac surgeons should continue to spearhead the efforts to investigate the role of hybrid interventions in the treatment of thoracic aortic disease.

Reference