Abnormalities of vertebral fusion branches, rather as a vessel segmentally fused to the subclavian artery and giving intercostal space. The left internal mammary artery could be seen as a vessel not originating from the second intercostal space.

3. Discussion

In this case, the LIMA was found to be originating from the 2nd intercostal space. The anatomy of the vessel, proximal to this, was unclear. The decision was taken to use it as a free graft as opposed to a pedicle as there was potential for a steal phenomenon to occur. This patient had Klippel-Feil syndrome, a rare congenital disorder characterised by congenital fusion of two or more cervical vertebrae and may be associated with other organ system anomalies [3]. There are no reports in the literature about coronary artery bypass grafting in such patients. There are some case reports highlighting the presence of subclavian artery anomalies in patients with Klippel-Feil syndrome [4, 5]. Bavinck and Weaver use the term subclavian artery supply disruption sequence (SASDS) and hypothesise that this is a common pathogenesis for a number of recognised birth defects. In a disruption sequence, a developmentally normal embryo or fetus experiences a destructive process, in this case partial or complete blockage of blood flow in the subclavian artery, with cascading consequences [5]. The exact nature of the defect will depend upon the exact site and timing of the disruption sequence. A number of similar defects have been described of which Klippel-Feil syndrome is well recognised. This theory relies in the subclavian artery and its branches develop by the process of sprouting angiogenesis.

More recent research in the zebrafish animal model suggests the vasculature in the trunk develops by the process termed vasculogenesis [6]. This is the process of blood vessel formation occurring by a de novo production of endothelial cells. Coalescence of these segmental angioblasts results in the formation of the trunk vessels. The left internal mammary artery could be seen as a vessel not ‘sprouting’ from the subclavian artery and giving intercostal branches, rather as a vessel segmentally fused to the subclavian artery. Abnormalities of vertebral fusion/segmentation may be linked to abnormal segmentation of the trunk vasculature thus explaining the phenotype seen in our patient. Evidence for faulty segmentation rather than fusion of the vertebrae is provided in a genetic study of Klippel-Feil syndrome families by Clarke et al. [7].

For these reasons, we propose that any patient with Klippel-Feil syndrome for coronary artery bypass grafting, where the use of the internal mammary artery is being considered for conduit, should have imaging of the subclavian and internal mammary arteries pre-operatively as there is likely to be an associated anomaly. This will allow better planning of the operation in terms of conduit choice.

References


ICVTS on-line discussion A

Title: Thoracic bifurcation and Klippel-Feil syndrome

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eComment: The article by Paul et al. [1] proposes that any patient with Klippel-Feil syndrome for CABG, should have imaging of the subclavian and internal mammary arteries preoperatively as there is likely to be an associated anomaly. My search of our database finds that we performed 2506 CEA procedures (last ten years), and one patient was followed up for a Klippel-Feil anomaly (plain radiography, CT scan, MRI, DSA, ultrasound of the kidneys, intravenous pyelography, hearing (loss evaluation). The patient presented with bilateral thoracic carotid bifurcation and the internal carotid artery (ICA) being anterior and medial to the external carotid artery (ECA) on both sides. Until today, the number of documented cases remains insufficient to draw a significant conclusion of association between the Klippel-Feil anomaly and subclavian artery supply disruption sequence (SASDS). It is generally accepted that the precursor of the CCA-ECA trunk arises from the aortic sac and secondarily migrates toward the third aortic arch to constitute carotid bifurcation. Alternatively, it has been proposed that the ventral pharyngeal artery arises directly from the third arch at the location of the future carotid bifurcation. In both cases, persistence of the duc tus caroticus associated with partial or complete involution of the third will increase the distance separating the CCA and ECA embryologic origins. Migration of the ECA toward the carotid axis then being incomplete or absent, resulting either in a low position of the carotid bifurcation or in separate origins of the ECA and ICA. The association of low carotid bifurcations with a Klippel-Feil anomaly is interesting in that it might point to a segmental developmental disorder leading both to the fusion of cervical vertebral bodies and to the third aortic arch anomaly facilitating SASDS. Intrathoracic carotid bifurcations may also represent diagnostic pitfalls by lying outside the region of interest during pre-operatively diagnostic evaluation. As anomalies of carotid bifurcation are joint with subclavian as well as internal mammary artery it could be wise to SASDS be represented by the above conditions. Since this is a syndrome with a constellation of possible abnormalities, no set of definitive contraindications for cardiovascular surgical subspecialty exists. If a surgeon believes that an operation is...
indicated, it is incumbent upon her/him to make certain none of the other conditions that could cause morbidity or mortality are present. A thorough workup of the patient (especially preoperative imaging techniques) is imperative prior to cardiovascular surgical intervention. Greater interest, observations, and endeavors in this field should offset the problems of diagnosis.

Reference