branches, rather as a vessel segmentally fused to the
blasts results in the formation of the trunk vessels. The
endothelial cells. Coalescence of these segmental angio-
vessel formation occurring by a de novo production of
gests the vasculature in the trunk develops by the process
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 subcla-
vian 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

syndrome – the risk of cervical spinal cord injury: a case report. BMC
syndrome and associated congenital abnormalities: evaluation of 23
876–879.
[4] Brill CB, Peyster RG, Keller MS, Galtman L. Isolation of the right
subclavian artery with subclavian steal in a child with Klippel-Feil
anomaly: an example of the subclavian artery supply disruption
[5] Bavinck JN, Weaver DD. Subclavian artery supply disruption sequence:
hypothesis of a vascular etiology for Poland, Klippel-Feil, and Mobius

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 subcla-
vian artery supply disruption sequence (SASDS) and hypothe-
sise that this is a common pathogenesis for a number of
recognised birth defects. In a disruption sequence, a devel-
opmentally normal embryo or fetus experiences a destruc-
tive process, in this case partial or complete blockage of
blood flow in the subclavian artery, with cascading conse-
quences [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 sug-
gests 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 angio-
blasts 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/seg-
mentation 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 subcla-
vian 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.

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syndrome – the risk of cervical spinal cord injury: a case report. BMC
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ICVTS on-line discussion A

Title: Thoracic bifurcation and Klippel-Feil syndrome
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doi:10.1510/icvts.2006.141994A

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 CCA 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
ductus 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 bifurca-
tions 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 evalu-
ation. As anomalies of carotid bifurcation are conjoint 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 cardiovas-
cular 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