of the pathogenesis of renal failure and could yield novel treatment strategies. Until these genes are identified, it would be prudent for physicians caring for hypertensive and diabetic patients to identify those having relatives with nephropathy. Hypertension, hyperlipidaemia and diabetes mellitus should be treated aggressively in these individuals at high risk of developing future renal disease.

References


Vascular steal syndrome and ischaemic monomelic neuropathy: two variants of upper limb ischaemia after haemodialysis vascular access surgery

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Introduction

Distal ischaemia following placement of upper limb arterio-venous accesses for haemodialysis occurs with widely varying frequency depending on surgical technique and experience, location and type of access created and the patient population. Incidence rates of ischaemic complications range from 0.24 to 42% [1,2] but are most often between 1 and 9% [3–6]. Vascular steal syndromes ranging in severity from mild digital paraesthesiae to catastrophic acute arterial insufficiency may occur. Ischaemia may also affect upper limb nerves predominantly or exclusively, and result in the under-recognized, but distinct clinical entity of ischaemic monomelic neuropathy.
Diabetic haemodialysis patients are at increased risk of upper limb ischaemia after access surgery because of often severe digital calcific atherosclerosis and pre-existing diabetic neuropathy. Accesses originating from the brachial artery are also a major predisposing factor, as this vessel constitutes the sole arterial inflow to the forearm and hand, and in the absence of collateral vessels about the elbow, diversion of all or most of high flow brachial arterial blood through an access will produce distal ischaemia.

**Vascular steal syndromes**

In mild cases of vascular steal, the onset is insidious and often delayed for days, weeks or months. Symptoms include numbness, painful paraesthesiae, stiffness and swelling of one or more fingers, and are sometimes precipitated by, or exacerbated during haemodialysis. Intra-dialytic lowering of systemic blood pressure or pO\(_2\) may explain the dialysis-associated symptom exacerbation [7]. The radial pulse is usually present. Symptoms may resolve spontaneously over weeks or months.

In more severe cases, numbness and pain progress and are associated with pallor, coolness, diminished sensation, ischaemic ulcers, trophic changes and progressive dry gangrene of one or more digits (Figure 1). The radial pulse is absent. In very severe cases of vascular steal, changes are apparent on the operating table with immediate pain, pallor, pulselessness, and paralysis of the hand.

The pathogenesis of vascular steal with a side-to-end radiocephalic fistula is shown in Figure 2. The low pressure run off system afforded by the fistula causes reversal of blood flow from digital and palmar arch arteries through the distal limb of the radial artery. In patients with pre-existing athero-occlusive narrowing of the palmar arch arteries and their tributaries, even mild degrees of flow reversal may result in clinically significant steal. In patients with normal palmar arch arteries, more severe degrees of flow reversal would be necessary to produce clinical symptoms. End-to-end rather than end- or side-to-side fistulae are therefore preferred in order to eliminate the distal arterial limb through which retrograde flow occurs. In the brachial location, end-arterial fistulae are not possible, hence the distal brachial arterial limb and its tributaries provide conduits for flow reversal.

Vascular steal syndromes can usually be diagnosed clinically. Cardinal findings are symptom relief and reappearance of the radial pulse with manual occlusion of the venous limb of the access (abrogating retrograde flow). When physical symptoms and signs are early, mild or atypical, and the diagnosis uncertain, Doppler ultrasound studies showing severe flow reversal and digital plethysmography (pulse volume recordings (PVRs)) documenting digital pressures < 50 mmHg, and digital-brachial indices < 0.47 [8] with symptom relief and augmentation of the pulse wave with fistula compression, are diagnostic. Fistulography is indicated in cases of severe hand ischaemia after access surgery, as it may show potentially remediable athero-occlusive or embolic arterial disease with or without the presence of retrograde arterial flow and coexisting clinical steal syndrome [9]. It is important to note that demonstration of retrograde arterial flow does not predict or...
indicate existence of a clinical steal syndrome. Retrograde blood flow is a physiologic consequence of the rheology of an arteriovenous access, and up to 67% of radiocephalic fistulae evaluated by extravascular electromagnetic flowmetry [10], and 86% of proximal arteriovenous grafts evaluated by digital plethysmography [8], show clinically silent retrograde flow. Digital pulse oximetry is also useful in diagnosing steal syndromes [5]: in 5 patients with side-to-side arteriovenous fistulae, symptoms suggestive of steal, yet normal physical examination, arterial oxygen saturations were low in all cases and rose to normal levels (>90%) with fistula compression.

**Treatment of vascular steal**

Severe cases of vascular steal require ligation or removal of the access. Ligation of the distal radial limb of a side-to-side radiocephalic fistula (Figure 2) is curative. Various techniques of fistula banding [4,11] may also be used, but there is risk of subsequent access thrombosis. Ligation of the arterial limb just distal to the access and placement of an interposition graft from the proximal arterial inflow to a more distal artery is technically demanding but often successful [4].

No reliable predictors of development of a steal syndrome exist. Allen’s test documents patency of the ulnar artery, and should be performed prior to placement of radiocephalic fistulae. The patient is instructed to clench and release his fist several times, then to make a tight fist so that venous blood is forced from the palm. The radial and ulnar arteries are manually occluded by the examiner until the palm blanches. The ulnar artery is released, and the palm should flush immediately if arterial inflow through this vessel is intact. The test however, is applicable only to radiocephalic fistulae, and may be falsely negative in the 9–20% of the population with aberrant upper limb arterial anatomy. Further pre- or intra-operative evaluation is warranted in patients deemed to be at high risk based on prior steal syndromes, known peripheral vascular disease, advanced age, or presence of diabetes. A systolic blood pressure difference in the upper limbs of >20 mmHg, and intra-operative digital-brachial indices <0.47, or digital pressures <50 mmHg are suggestive, but not absolutely predictive of vascular steal [8]. Intraoperative monitoring of digital oxygen saturation may prove useful in guiding the anastomotic size of arteriovenous fistulae and graft, and in banding operations [12].

**Ischaemic monomelic neuropathy**

Ischaemic neuropathy of upper limb nerves after dialysis access surgery was first reported by Bolton in 1979 [13]. A more detailed description of the condition and coining of the term ischaemic monomelic neuropathy (IMN) came in 1983 [14]. Subsequent reports have remained confined to neurologic and surgical literature [15–17] so that under- and mis-diagnosis of IMN is frequent in the realm of renal medicine. The condition is rare, and precise incidence figures are not available from existing reports. IMN is seen almost exclusively in diabetic haemodialysis patients [18], particularly older ones, with pre-existing peripheral neuropathy and/or peripheral vascular disease. Acute pain, weakness and paralysis of the muscles of the forearm and hand (often with prominent sensory loss and dysesthesiae) occur immediately (within minutes to hours) of placement of an arteriovenous access in the brachiocephalic or antecubital location. The condition is not seen with accesses originating distal to the brachial artery. IMN results from sudden diversion or transient occlusion of the blood supply to the nerves of the forearm and hand, the acute ischaemic insult being severe enough to damage nerve fibres, but insufficient to produce necrosis of other tissues. The condition may therefore be most simply described as a steal syndrome affecting only the nerves. This selective neural injury may be due to the greater metabolic requirements and more tenuous blood supply of peripheral nerves when compared to other tissues [19]. The antecubital area may also be the ‘watershed area’ for the vasae nervorum of the three upper limb nerves [20].

IMN can be diagnosed clinically based on immediate symptom onset and dominant neurologic symptoms and signs. The hand is warm, though the radial pulse is variably present. There are no signs of muscle infarction such as tenderness or pain with passive extension; and signs of ongoing vascular insufficiency and trophic changes are usually absent. Features differentiating IMN from vascular steal syndrome are shown in Table 1. Nerve conduction studies show axonal loss and reduced sensory and motor nerve conduction velocities of median, radial and ulnar nerves [12]. One study reports a predilection for earlier and more severe (but not isolated) median nerve involvement [21]. Electromyography reveals severe acute denervation of all upper limb nerves which is maximal distally [13,16]. Digital pressures are >50 mmHg and digital-brachial pressure indices >0.3

| Table 1. Differentiating vascular steal syndrome from ischemic monomelic neuropathy |
|-------------------------------------|------------------|
| Steal syndrome                      | IMN               |
| Onset                               | usually insidious |
| Predilection for diabetes           | acute            |
| Access location                     | wrist, forearm, upper arm |
| Tissue affected                     | skin > muscle > nerve |
| Degree of ischaemia                 | severe, diffuse   |
| Radial pulse present                | + + + +           |
| Digital pressures                   | mild-moderate     |
| Reversibility                       | + + + +           |

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Differential diagnosis of IMN

IMN may be misdiagnosed as a complication of axillary block or patient positioning during surgery; or attributed to post-operative or psychogenic pain. Involvement of a single upper limb nerve excludes the diagnosis of IMN. Paralysis of a single nerve in the setting of vascular access surgery should prompt a search for local nerve compression secondary to haematoma, aneurysm or abscess [22]. Some cases of carpal tunnel syndrome may also manifest or exacerbate following access surgery, perhaps related to oedema and venous hypertension in the area of the ﬂexor retinaculum, or to a component of vascular steal [23,24]. It is unlikely that cases of ipsilateral carpal tunnel syndrome after access surgery are related solely to the access, either via ischaemic neuropathy or steal syndrome. In early reports of CTS in association with haemodialysis accesses, the onset of CTS was one or more years after access surgery [25,26], and retrospective staining of transverse carpal ligament biopsies revealed beta-2-microglobulin amyloid in half of the cases in one report [25].

Treatment of IMN

Immediate closure of the access is required upon diagnosing IMN in order to prevent severe and irreversible neurologic injury. Even with early access closure, paralysis and pain may be permanent or only partially reversible [15–17]. Delay in diagnosis will of course reduce the likelihood of improvement, hence recognition of this uncommon complication of vascular access surgery by surgeons and nephrologists is crucial.

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[16]. Fistulography is indicated in cases where there is clinical uncertainty and in search of correctable embolic or athero-oclusive disease of the inﬂow or distal arteries [9].