

Superior vena cava syndrome: anaesthesia for renal access

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The incidence of end-stage renal disease (ESRD) is increasing exponentially, in parallel with the prevalence of diabetes, hypertension and obesity. The inevitable consequence is a dramatic increase in demand for dialysis and transplantation.

Patients requiring urgent renal dialysis access are among the most sick who present for anaesthesia. When superior vena cava syndrome (SVCS) is superimposed, further anaesthetic challenges arise.

Issues to consider in such a patient include:

- The pathophysiology of the condition underlying the ESRD
- Pathophysiological consequences of the ESRD itself
- Consequences of chronic medications in ESRD
- Further implications of a period of failed, or inadequate, dialysis, as a consequence of inadequate access (the indication for the access procedure)
- The pathophysiological effects of SVCS
- Surgical considerations
 - Potential vascular access sites
 - Stenting requirement
 - Potential complications of difficult access
- Choice of anaesthesia.

The scope of this talk does not allow for consideration of the all of the pathophysiological implications of diabetes and hypertension. However, the other issues will be examined.

Pathophysiology of end-stage renal disease

Patients with ESRD encounter problems relating to:

- End-organ damage from the underlying cause of ESRD
- End-organ damage from the pathophysiological consequences of the ESRD itself
- Chronic drug therapy

- Advancing age [renal transplantation (RT) now regularly performed in over-60s].

Table I contains an incomplete list of the pathophysiological consequences of ESRD, some of which require further discussion.

Cardiac and cardiovascular disease is the norm in ESRD (92% of patients). Its incidence is 10-30 times greater than a population with normal renal function. RT reduces the incidence of cardiac disease to only twice that of the normal population. Cardiac deaths account for > 50% of ESRD mortalities, and 50% of post-transplant deaths. The age-adjusted incidence of cardiac mortality in ESRD is 700 times that of the non-renal failure population. Myocardial infarction in the ESRD population has a poor prognosis, with a 40% one-year and 60% five-year mortality rate. RT reduces the risk of cardiac death 70-fold.

RT has been shown to reduce the incidence of all adverse effects of ESRD markedly. This is most evident when a transplant occurs early after commencement of dialysis. Best recipient and graft outcomes are observed if RT is performed within a year of the onset of dialysis. Outcomes of RT after < 6 months of dialysis are similar to those of pre-dialysis transplantation.

Cardiovascular system (CVS) pathophysiological consequences of ESRD include:

- Volume overload
- Left ventricular (LV) hypertrophy/dilatation/dysfunction
- Accelerated coronary artery disease
- Accelerated cerebrovascular disease.

The aetiologies of the above are multifactorial, and include:

- Fluid retention
- Underlying and secondary hypertension
- Anaemia

Table 1: Pathophysiological consequences of chronic renal failure important to anaesthetists

Abnormality	Clinical presentation or mechanism of abnormality
Arterial hypertension	Left ventricular hypertrophy, hypertensive cardiomyopathy, malignant hypertension
Cardiovascular abnormalities	Congestive heart failure, uraemic pericarditis, cardiac tamponade, dysrhythmias (K ⁺ , Ca ²⁺)
Anaemia	Decreased erythropoietin production, diminished erythrocyte survival (normochromic, normocytic anaemia)
Platelet dysfunction	Bleeding diathesis
Electrolyte imbalance	Hyperkalaemia, hypocalcaemia, hypermagnesaemia
Fluid disturbances	Hypervolaemia (hypertension, pleural effusions, congestive heart failure, pulmonary oedema), intravascular volume depletion post-dialysis
Acid-base abnormalities	Metabolic acidosis
Gastrointestinal abnormalities	Uraemic gastroenteritis, nausea and vomiting, peptic ulcer, bleeding, reflux
Endocrine abnormalities	Secondary hyperparathyroidism, osteomalacia
Dialysis-related problems	Peritonitis (peritoneal dialysis), systemic anticoagulation, vascular access, dementia, disequilibrium syndrome after acute dialysis (cerebral oedema)
Central nervous system dysfunction	Convulsions, myoclonus, irritability, lethargy

- Dyslipidaemia
- Hyperhomocysteinaemia
- Hyperphosphataemia and calcium/phosphate overload
- Micro-inflammation [one-third to two-thirds have elevated C-reactive protein (CRP)]: endothelial dysfunction underlies much of the CVS pathophysiology in ESRD
- Duration of uraemia and dialysis.

The major factors that provoke ventricular hypertrophy, dilatation, wall movement abnormalities and dysfunction are anaemia, hypertension and volume overload. These are ameliorated by effective dialysis and antihypertensive or diuretic therapy. Significant hypertension more than doubles mortality in dialysis patients, and mortality rate is further doubled for every 10 mmHg further rise in mean arterial blood pressure (MABP). As mentioned, RT can result in restoration of ventricular anatomy and function, provided it follows soon after commencement of dialysis. A delay of longer than 3 years between onset of dialysis and RT is associated with a degree of cardiac myocyte necrosis, and failure to restore ventricular function fully. The impact of anaemia on LV function is a composite of the reduced rheology with consequent high output state, tachycardia with reduced diastolic duration, and reduced oxygen-carrying capacity. Correction of anaemia, using recombinant erythropoietin (EPO), to a haemoglobin (Hb) of 10-12 g/dl, but not > 12 g/dl, produces an average 60% improvement in ventricular function. Further improvement requires blood pressure control and effective volume and electrolyte control with dialysis.

There is an 18% increase in mortality for every g/dl reduction in Hb below 10 g/dl, and an unspecified increase in risk of ischaemia if Hb rises above 12 g/dl. RT produces

EPO alpha-free status in 80% of recipients. Correction of anaemia with EPO, in addition to improving O₂ supply and reducing ventricular hypertrophy and dysfunction, improves functional capacity and quality of life, as well as coagulation status and platelet function. However, it may produce or compound hypertension.

Long-term ESRD patients have > 90% incidence of coronary artery disease. This is consequent, in part, on the underlying diabetes or hypertension. However, numerous associations or consequences of ESRD may accelerate the process.

These include:

- Anaemia
- Fluid overload and hypertension
- Tachycardia and reduced diastolic myocardial perfusion time
- Increased ventricular thickness and wall tension
- Dyslipidaemia
- Chronic inflammatory states
- Hyperhomocysteinaemia is an independent predictor of CVS mortality in ESRD patients, and if sustained, of morbidity post-RT. It is also thought to promote coronary calcification.
- Ca/PO₄ overload with heterotopic coronary calcification. Secondary hyperparathyroidism and a raised Ca-PO₄ product can produce valvular and coronary calcification. Increased myocardial Ca content predisposes to LV hypertrophy and dysfunction. Ca-based phosphate chelators may lead to hypercalcaemia, and an identical picture.
- Elevated levels of asymmetric dimethylarginine (ADMA), a nitric oxide (NO) antagonist, are seen in ESRD

Other pathophysiological observations include:

- Pulmonary hypertension, probably thromboembolic in origin, is present in 40% of haemodialysis, but not peritoneal dialysis, patients. It reverses after successful RT.
- Coagulation disorders include:
 - Impaired platelet function, inversely proportional to the quality of the dialysis
 - Reduced levels of Factor VIII and Von Willebrand's factor
 - Systemic heparinisation, related to haemodialysis
 - Hypercoagulability related to underlying disease, e.g. systemic lupus erythematosus (SLE).
- Hyperkalaemia. Patients appear to develop a new potassium set point, and are better able to tolerate K levels of 5-6 mmol/l.
- Hepatitis C may cause ESRD (via cryoglobulinaemia and membranoproliferative glomerulonephritis), and may complicate dialysis. It is an independent predictor of worse outcome of dialysis, and poorer prognosis of the transplanted kidney.
- Altered pharmacokinetics (PK) and dynamics (PD) may affect choice and effects of drugs chosen for anaesthesia and therapy.

Chronic medications in chronic renal failure

ESRD patients are generally on a melange of drugs to control underlying disease, and the pathophysiological consequences of the ESRD. These include insulin, oral hypoglycaemic agents, angiotensin-converting enzyme (ACE) inhibitors (especially in type 1 diabetics), angiotensin-receptor blockers (ARBs) (especially in type 2 diabetics), β blockers, calcium-channel blockers, diuretics, statins, antacids, proton-pump inhibitors, laxatives, iron, vitamin supplements, recombinant EPO, vitamin D analogues, phosphate binders, aspirin, and warfarin [in patients with synthetic arteriovenous (AV) fistulae]. As a general rule, medications should be continued up to and including the morning of surgery, with the usual perioperative provisos in respect of antidiabetic agents, aspirin and warfarin.

It is crucial that antihypertensive agents, particularly β blockers and calcium-channel antagonists, are administered on the day of surgery to help mitigate the almost inevitable rebound hypertension, tachycardia, and myocardial ischaemia seen after surgery. Moderate hypertension is not regarded as a contraindication to surgery, but does predict intraoperative hypotension and postoperative hypertension, and provision must be made for their management.

ACE inhibitors have been shown to reduce mortality in congestive cardiac failure. In addition, they are known to slow the progression of the nephropathy in a variety of nephritides. ACE inhibitors and ARBs can reverse left ventricular hypertrophy (LVH), and improve lusitropy in transplant recipients, independent of anti-hypertensive effect. However, it is advisable to administer the last dose of such agents before the last session of dialysis (ideally the afternoon before surgery). This will limit the profound hypotension observed when anaesthetising such patients after a same-day dose of drug, particularly against the background of intravascular volume depletion from recent dialysis. It will also facilitate the resuscitation of patients with perioperative surgical bleeding.

In the emergency setting, we rarely have any control over compliance with, and timing of, chronic medications.

Implications of inadequate dialysis

Inadequate dialysis in ESRD patients produces exacerbation of all of the pathophysiological effects of uraemia, and may present like acute renal failure superimposed on ESRD, with:

- Severe volume overload (exacerbating impact of SVCS)
- Severe acidosis
- Severe electrolyte imbalance, and notably hyperkalaemia, hyperphosphataemia, and hypocalcaemia with consequences in respect of CVS and central nervous system (CNS) function
- Severe platelet dysfunction and increased bleeding potential; worsened by the hydrostatic effects of SVCS
- Other uraemic effects: Gastroparesis, CNS depression, and worsening of PK abnormalities.

Pathophysiological effects of superior vena cava syndrome

SVCS occurs as a result of partial, or complete, obstruction of SVC flow, as a consequence of:

- Compromised vessel anatomy, usually tumours
- Compromised vessel wall integrity
- Compromised venous flow.

SVC vascular access devices, particularly indwelling ones, may produce SVCS, based on the latter two mechanisms. In 2010, 40% of reported cases related to intravascular devices, reflecting the massive increase in utilisation.

The severity of SVCS depends on:

- Degree of obstruction.
- Level of obstruction. This is worse if below the azygos confluence.
- Rate of evolution of obstruction. Collaterals require a few weeks to form and accommodate the increased venous volume above the obstruction.

Complications of SVCS relate to:

- Increased hydrostatic pressure above the obstruction, leading to oedema or effusions throughout the respiratory tract and in the CNS.
- Dilated collateral veins (azygos, intercostals, paravertebral, thoracoacromial, and chest wall). Sometimes, this is not clinically obvious.
- The risk of venous thromboembolism (VTE) consequent upon catheter-related thrombosis. Thrombosis occurs in around 45% of central vascular devices [pulmonary embolism (PE) in 12%].
- The risks of anticoagulation for thrombosis, especially on the background of platelet dysfunction (~ 6% risk of major bleeding).
- The severity of SVCS is graded 0 (asymptomatic) to 5 (fatal), depending on respiratory and neurological signs and symptoms using the Yale modification of the Kishi Scoring System (Table II).

Features of imminent CNS and or respiratory compromise mandate urgent imaging, and probably invasive intervention, e.g. balloon angioplasty or stenting.

Surgical considerations

Potential sites for vascular access

- Complete obstruction rules out head, neck, and upper limbs as access sites.

Table II: The Kishi Scoring System for signs and symptoms of superior vena cava syndrome

Signs and symptoms	Grade
Neurological symptoms	
Stupor, coma or blackout	4
Blurry vision, headache, dizziness, or amnesia	3
Changes in mentation	2
Uneasiness	1
Laryngopharyngeal or thoracic symptoms	
Orthopnoea or laryngeal oedema	3
Stridor, hoarseness, dysphagia, glossal oedema, or dyspnoea	2
Cough or pleural effusions	1
Nasal and facial signs or symptoms	
Lip oedema, nasal stiffness, epistaxis, or rhinorrhoea	2
Facial swelling	1
Venous dilatation	
Neck vein or arm vein distension, upper extremity swelling, or upper body plethora	1

- Partial obstruction: The safety of further cannulation has not been established.
- Suspicion of SVCS mandates imaging to delineate the nature and extent of the obstruction.

Requirements for stenting

- Based on clinical features and imaging
- Range of interventions: Catheter-directed thrombolysis, balloon angioplasty, and stent placement (requiring long-term anti-platelet therapy)
- Usually under local anaesthetic, via intravenous or subclavian vein
- Outcome of stenting: 50-60% three-year patency
- Permits use of neck or upper limbs for access within a few weeks of correction of the obstruction.

Potential complications of difficult access

- Pneumothorax
- Major vascular injury with catastrophic bleeding, necessitating thoracotomy
- Air embolism
- VTE
- Sepsis.

Choice of anaesthesia

Intravenous (IV) access for induction must be in the lower limbs in the case of complete SVC obstruction. With partial obstruction, upper limb access is acceptable, if not desirable. Collateral venous drainage means induction time will be prolonged, and overdose is a potential risk. Vascular access must be of adequate size to cope with substantial blood loss in the event of a major vascular injury. Again, upper limb access will result in delays in resuscitation fluids and drugs reaching the central circulation. In the absence of major bleeding, fluid restriction is the watchword. Crystalloids are appropriate, although saline may worsen acidosis if administered in substantial volumes.

If the dialysis access is to be via upper limb or groin, arteriovenous fistula (AV) fistula or graft, or a femoral dialysis catheter, then unsedated local or regional anaesthesia are excellent options, avoiding many of the major anaesthetic challenges:

- Profound sensitivity to CNS depressants (cerebral oedema from SVCS and altered PK of drugs in ESRD).
- Prolonged neuromuscular blockade (delayed renal excretion and PK changes).
- Difficult airway (glottal and laryngeal oedema, and risk of haemorrhage).
- Worsening hypoxia due to general anaesthetic-related atelectasis, in the presence of fluid overload from inadequate dialysis and fluid transudation from bronchial venous (collateral) hypertension.

- The catastrophic dynamic airway collapse, seen with superior and anterior mediastinal tumours, should not occur here.
- Reflux and aspiration (emergency: unknown fasting status, and uraemic gastropathy).
- Haemodynamic instability. Intubation hypertension, intraoperative hypotension and postoperative hypertension are typical in ESRD patients on antihypertensives. This is also the case with fluid sequestration in the head, neck and torso of SVCS patients in the supine position. There is a greater risk of myocardial ischaemia and ventricular dysfunction, especially if overloaded from inadequate dialysis. There is potential for massive bleeding from major vascular injury in the presence of distorted anatomy, from previous vascular access points, and increased hydrostatic pressure in SVCS.
- Risks related to electrolyte disturbances, especially hypokalaemia, hyperphosphataemia, and hypocalcaemia: Dysrhythmias, myocardial ischaemia, muscle weakness, prolonged neuromuscular blockade and CNS irritability.
- Excessive bleeding: Coagulopathic (platelet dysfunction, and heparin from dialysis), hydrostatic or surgical.
- Potential for spinal and bone injuries with uraemic osteoporosis and malacia.
- Development of a major surgical complication: Pneumothorax, vascular injury, and air embolism.
- Contrast-related complications, where radiography is required.

- Delayed awakening for many of the above reasons in the face of a short surgical procedure.

Head and neck vascular access procedures are feasible under local anaesthetic, but the comfort and safety of these profoundly obtunded patients may be compromised. Brief general anaesthesia with intubation, with controlled ventilation and adequate peripheral IV access, is preferable.

My technique of choice is:

- Achieve lower-limb vascular access.
- Proper preoxygenation: often head-elevated position.
- Rapid sequence induction with a reverse oral RAE tube one size smaller than usual, using:
 - Propofol 1.5 mg/kg mixed with 20 mg of etilefrine/ephedrine over 20 seconds.
 - Remifentanil 3-5 µg/kg over 10 seconds, immediately thereafter.
 - 20 ml flush solution to hasten the onset of the above.
 - No manual ventilation.
 - Intubate when heart rate drops by 10%.
 - No relaxants.
- Ventilate with > 5 cmH₂O of positive end-expiratory pressure (PEEP) with sevoflurane or desflurane, and commence a remifentanil infusion if necessary.
- Awake extubation, head up.
- Postoperative chest X-ray.