

Anaesthesia in connective tissue disorders

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Summary

Patients with the more common connective tissue disorders require surgical operations more frequently than has been realized. They may present the anaesthetist with many potential problems. A few minutes of careful questioning and examination pre-operatively may prevent a tragic situation. A history of drug therapy is essential pre-operative information, particularly since many of these patients will need augmentation or coverage with steroid drugs. The anaesthetist must be aware of the patient's general state of health and must search for evidence of pulmonary, cardiac or haematological abnormalities.

S Afr Med J 1985; 67: 336-339.

Connective tissue varies from the loose skin below the eye to dense tendon, and consists of three components: cells, fibres and ground substances. All three components are under the control of physiological and hormonal mechanisms.

Disorders of the connective tissue elements themselves comprise two contrasting groups, the very common degenerative diseases and the rare inherited connective tissue disorders (osteogenesis imperfecta, Marfan's syndrome). With the exception of rheumatoid arthritis (a disease affecting perhaps 3% of the population), the connective tissue disorders are rare.

Patients are systemically and often chronically ill, and conditions such as anaemia, hypovolaemia and hypoproteinaemia may influence the anaesthetic technique. Many patients, for instance, have pulmonary manifestations of their disease, which puts them at added risk. The usual precautions apply to patients who are receiving, or who have recently received, corticosteroids; their adrenocortical reserve may be sufficient for the ordinary needs of life but inadequate to meet the extra burden of anaesthesia and operation. As short a course as 1 week may depress cortical function, and in some cases of prolonged therapy depression may last as long as 1-6 years.

There are no satisfactory simple tests for adrenocortical reserve,¹ and the tests for pituitary-adrenal axis function are too complex and expensive for routine use. In clinical practice it must therefore be realized that despite the fact that in some patients on maintenance levels of steroids activation of the pituitary-adrenal axis still occurs in response to stress, we cannot easily predict whether or not this will take place. We are left with an obligation to provide steroid coverage for patients undergoing surgical stress regardless of the magnitude

of the procedure; adrenal insufficiency has been reported following a simple bunionectomy.

Collapse is unlikely to occur more than 2-6 months after cessation of treatment, but it is generally safer to give corticosteroid cover in the form of intravenous or intramuscular hydrocortisone than to omit it in cases of doubt. Corticosteroid administration can be commenced at the time of premedication and can be continued for 3 days after major surgery, for 24 hours following minor operations, or it can be restricted to a single injection before endoscopy or other brief procedures. Hydrocortisone hemisuccinate can be given (to an adult) in 100 mg doses 6-hourly. One of several protocols for administering prophylactic pre-operative cortisone acetate may be used.²

Rheumatoid arthritis (RA)

RA most often starts between the ages of 25 and 55 years, affecting women about three times as often as men. The current view on the aetiology of RA involves the trio of infection, hypersensitivity and auto-immunity.³

The anaesthetist is likely to see patients requiring surgery for corrective orthopaedic procedures and for complications of corticosteroid therapy. Optimal anaesthetic management should begin with a detailed pre-operative assessment and preparation. This same degree of attention and expert care must then be carried into the peri- and postoperative periods. The pre-operative checklist in Table I is recommended.⁴

TABLE I. PRE-OPERATIVE CHECKLIST IN RA

Anaesthetic history
Drug history — the possibility of drug interactions with the anaesthetic agents must always be kept in mind²
Examination of neck and jaw mobility
Indirect laryngoscopy, if indicated
Lung function tests, including blood gas analysis
Chest radiographs; skeletal radiographs, if there is limitation of spinal movement
ECG
Blood profile, including determination of haemoglobin value, ESR, white cell count and platelet count
Urinalysis
Determination of creatinine clearance rate and test for occult blood in stools if indicated

For general anaesthesia tracheal intubation should be performed, since reliance on mask anaesthesia may result in loss of patency of the airway. We would advise caution when administering premedicant drugs, which can increase pre-existing respiratory obstruction. Obstruction of the airway is a real problem in patients with RA. The crico-arytenoid joints may be involved in as many as 26% of these patients; the glottic opening may be narrowed, but the most common airway problem is a flexion deformity of the cervical spine. The head and neck should be manipulated with great care during positioning or intubation, because cervical vertebral erosion and subluxation may occur especially at the atlanto-

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axial joint. Involvement of the temporomandibular joints may make visualization of the larynx difficult, and nasal intubation is often the method of choice.

As in all situations in which the patency of the upper airway is under suspicion, intravenous induction of anaesthesia should be used with care, and muscle paralysis is contraindicated unless it is established that the patient's lungs can be ventilated adequately. If there is doubt concerning the safety of the airway in the unconscious state, intubation under local anaesthesia with the patient awake may be necessary.⁵ A pre-operative tracheostomy may be indicated, particularly for major surgery. Additional problems arise in the patient with a full stomach. Here the airway must be secured as soon after loss of laryngeal reflexes as possible. This might be impossible when a difficult intubation is anticipated. It has been suggested in the literature that intubation under local anaesthesia with the patient awake guarantees protection of the airway in patients at an increased risk of regurgitation. The basis for this view is questionable.

Arterial blood gases should be measured pre-operatively in patients whose respiratory function may be affected adversely by the surgical procedure. Costochondral involvement may cause a restrictive defect, reduced vital capacity and low total lung volume, possibly accentuated by loss of compliance due to a specific pulmonary fibrosis. Ventilation-perfusion inequalities are common, leading to arterial hypoxaemia. It may be necessary to assist ventilation during general anaesthesia in patients who have rheumatoid lung changes and possibly costochondral involvement which limits chest wall expansion. It has been stated that patients with RA are extremely sensitive to agents which depress respiratory function.⁶ There is general agreement that patients with rheumatoid arthritis should not be left alone postoperatively and that it is wise to watch them carefully and even to assist respiration.

The pericardium, myocardium and endocardium can all be involved in the rheumatoid process and dysrhythmias may appear during anaesthesia. Monitoring is mandatory during anaesthesia. The most frequent cardiac signs and symptoms of RA are enlargement of the left ventricle, congestive heart failure and angina pectoris.

The presence of anaemia may mean that there is also hypovolaemia and hypoproteinaemia. Anaemia may be improved by transfusion of packed red cells before major surgery to raise the haemoglobin value to at least 10 g/dl. Oxygen flux to the tissues is further aided by maintaining cardiac output and attaining an optimal haemoglobin-oxygen dissociation curve.

Amyloidosis is a common renal problem in patients with RA. Renal involvement can reduce the excretion of drugs administered.⁴

The usual precautions apply to patients who are receiving, or who have recently received, corticosteroids. Most anaesthetic agents are safe, but the dosage must be moderated in the presence of muscle loss and hypoproteinaemia.

Postoperative problems are principally those involving lung function, and assisted ventilation may be required. Close supervision postoperatively with particular attention to the adequacy of respiration is essential. Infection and atelectasis are the most common postoperative complications. Cruickshank⁷ has indicated that the tendency for interstitial pneumonia to develop in the presence of RA is sufficiently pronounced for it to be regarded as a characteristic of the disease. The dosage of drugs which depress respiration should be controlled carefully.

In caring for the RA patient it must be remembered that no single protocol can be expected to manage all patients. Selection of an anaesthetic agent or agents does not appear to be a major factor in the safe outcome of a surgical procedure. No agent is categorically contraindicated and none is specifically beneficial

for the patient with RA. The choice of anaesthetic agents depends entirely on the extent of organ involvement and the experience and preferences of the anaesthetist. It is based on the type of surgery, the medical status of the patient and the surgical risks. Constant monitoring is the key to optimal management. An anaesthetic sequence which ensures the early return of consciousness and airway reflexes is recommended.

The same general considerations should govern the management of the associated or rheumatoid diseases, including Still's disease (juvenile chronic polyarthritis).

Ankylosing spondylitis (AS)

Cervical spine involvement is the lesion of most significance to the anaesthetist concerned with the management of the airway in patients with AS. There may be limitation of neck movement, which can cause intubation difficulties; complete ankylosis may be present. The risk of sustaining a cervical fracture may be increased in a small group of patients. The anaesthetic should be carefully planned in advance.

An important consequence of this disease of the spine is limitation of chest expansion by costovertebral joint involvement, so that breathing becomes predominantly abdominal. The impairment of rotation and limitation of spinal flexion can also significantly restrict chest expansion. The principal disability in patients with AS, however, is a restrictive pattern of lung function. Pre-operative respiratory assessment is essential and postoperative ventilation may be required after surgery. It is important to diagnose and treat pulmonary complications early, since they may occur in conjunction with limited chest expansion. An anaesthetic technique involving controlled ventilation may be preferred. Spinal or epidural anaesthesia may be difficult if not impossible if the intervertebral ligaments are calcified.

Also noteworthy is the association of aortitis, mitral insufficiency, conduction defects and aortic insufficiency.⁴ Patients may require a temporary pacemaker before surgery. Because it is of particular importance to avoid hypotension, constant monitoring during the operative procedure is needed.

Reiter's syndrome presents the same problems as other types of arthritis, but Sjögren's syndrome (keratoconjunctivitis, xerostomia and rheumatoid arthritis), with its loss of lacrimal and salivary secretions, may be an indication to omit the use of drying agents before and during anaesthesia. A rebreathing system is helpful in order to avoid excessive drying of the airway during endotracheal anaesthesia.

Approximately 25% of patients with agammaglobulinaemia develop a form of arthritis with many features of RA. Patients presenting for anaesthesia may be on corticosteroids.

Collagen vascular disorders

The name is merely a convenient label for a group of diseases; collagen is involved prominently in only one of them — scleroderma.

Systemic lupus erythematosus (SLE)

The aetiology of SLE is not apparent in most cases, but there is growing awareness that a number of drugs (e.g. hydralazine, isoniazid, para-aminosalicylic acid and procainamide) can cause the SLE syndrome. Renal involvement occurs in 75% of cases and over half the patients have involvement of the pericardium, endocardium or myocardium, while pulmonary infiltrates appear in many cases. The lines of treatment available include corticosteroids and cytotoxic drugs.

The cutaneous lesions may make fitting a face-mask difficult. Intubation may be necessary, but care is required since there are reports of crico-arytenoid arthritis in SLE. The patient may be very ill and febrile and the anaesthetist must be made aware of any renal, pulmonary or cardiac involvement. Anaemia is also very common and occasionally a haemorrhagic disorder may develop. In the presence of advanced renal disease the anaesthetist must carefully consider the need to give drugs primarily excreted by the kidneys.⁸ Adequate pre-operative and operative coverage with corticosteroids is required.

Scleroderma

Scleroderma is a multisystem disease and can present various anaesthetic problems.⁹ Sclerodermatous skin contractures can limit opening of the mouth, making conventional intubation difficult or impossible. Anatomical deformities may also make mask anaesthesia difficult. Patients may bleed profusely if traumatized during placement of the endotracheal tube. Inhalational induction may be difficult and potentially hazardous because of the problems of venous access and maintenance of an adequate airway. It is especially important in these patients to establish reliable intravenous access before induction of anaesthesia; venous cut-down or central venous catheterization may be necessary. Vasoconstriction may also interfere with blood pressure monitoring by the usual methods. Catheterization of the smaller peripheral arteries has been associated with spasm and subsequent necrosis; these vessels should therefore be avoided. The anaesthetic risk to patients with scleroderma may be increased because of involvement of various organ systems; pulmonary hypertension, renal lesions and episodes of left-sided heart failure have sometimes caused death. Sclerodermatous changes in the gastro-intestinal tract may result in malabsorption of vitamin K and thus lead indirectly to a blood-clotting abnormality.

Pregnancy accelerates the progression of scleroderma in 50% of patients. If regional anaesthesia is used for labour and delivery, smaller doses than usual of local anaesthetic agents are recommended because sclerodermatous patients may exhibit prolonged sensory and motor blockade afterwards. Although regional anaesthesia would obviate the difficulties and risks associated with endotracheal intubation and general anaesthesia, this may not be the safest approach.

The risk of aspiration of gastric contents may be compounded by the presence of gastro-oesophageal sphincter incompetence. Hypoxaemia resulting from decreased diffusion capacity is not unusual. Careful pre-operative assessment and a thorough understanding of the pathophysiological interactions are essential in formulating an anaesthetic plan.

Polyarteritis

This term is used to cover three overlapping conditions, which may even be variants of the same disease — polyarteritis nodosa, polymyalgia rheumatica and giant-cell arteritis. Many patients who develop polyarteritis already suffer from a chronic or acute respiratory infection.⁴ Renal lesions are frequent and may lead to renal failure or hypertension. The central nervous system may be involved, and lesions of the coronary vessels may cause myocardial infarction. Pulmonary lesions usually precede polyarteritis in other areas. Other common manifestations of this disease are joint lesions, sometimes causing chronic deformation of joints. Corticosteroids will certainly have been administered.

It is of interest to the anaesthetist that acute pharyngeal oedema and severe swelling of the uvula and parapharyngeal areas have been observed in patients with polyarteritis. Hypertension is invariable, and coronary and cerebral thrombosis is

then of major concern. If there is a suggestion of pulmonary involvement, any degree of lung dysfunction should be determined by appropriate tests. Any degree of renal failure should also be known pre-operatively.

Dermatomyositis/polymyositis

The hallmarks of this disease are oedema, dermatitis and multiple muscle inflammation. Three types of pulmonary involvement have been reported: (i) aspiration pneumonia, related to weakness of the muscles involved in swallowing; (ii) respiratory insufficiency resulting from progressive weakening of the intercostal muscles and diaphragmatic muscle; and (iii) lung involvement from the connective tissue itself, described as a patchy infiltrative process throughout both lungs. Involvement of the intercostal muscles often leads to severe pulmonary infection and is associated with a high mortality rate. Assisted ventilation should be considered early in patients with evidence of progressive ventilatory failure, and may need to be maintained for long periods.

Dermatomyositis is a systemic disease and the anaesthetist should be concerned with anaemia as well as intercurrent infection. Patients with dermatomyositis should require a smaller dose of a neuromuscular relaxant because of their diminished muscle mass. Ventilatory adequacy should be determined pre-operatively.¹⁰ Certain patients with this disease show a profound peripheral weakness, sometimes improved by administration of an anticholinesterase drug.

Muscle biopsy remains the most specific diagnostic investigation, but because of the frequency of patchy muscle movement, a negative biopsy does not exclude active disease. We would like to put in a word of caution about the routine use of general anaesthesia for the performance of muscle biopsies. While this may be necessary in some instances, especially in paediatrics, it is usually unnecessary and undesirable to 'anaesthetize the entire patient', especially if he or she has involvement of the respiratory muscles, for relatively limited surgery. Local anaesthetic drugs should not be injected directly into the muscle because of resulting histological changes but can well be used to perform field blocks. In addition, if muscle biopsies are being obtained for biochemical analysis it must be borne in mind that general anaesthesia can alter the normal biochemistry of muscles as well as of other tissues.

The first-line treatment of dermatomyositis still consists of systemic corticosteroids. For those patients who do not respond to steroids a variety of alternative treatments have been proposed, ranging from cytotoxic agents to total-body irradiation. Plasmapheresis has been used and often leads to removal of serum cholinesterase faster than it can be replaced by hepatic synthesis. The main clinical significance of serum cholinesterase is that it hydrolyses exogenous esters used in anaesthesiology (such as suxamethonium, procaine, chlorprocaine, tetracaine and propanidid) and thereby inactivates them.¹¹ Since serum cholinesterase is responsible for the biotransformation of suxamethonium, the reduction in its activity resulting from plasmapheresis must be seriously considered as a cause of prolonged relaxation when patients are treated with muscle relaxants of the succinylcholine type.

Granulomatous diseases

Wegener's granulomatosis

Wegener's granulomatosis is a multisystem disease which can involve any of the organs of the body and causes a constant triad of necrotizing giant-cell granulomatosis of the upper respiratory tract and lungs, widespread necrotizing vasculitis of the small arteries and veins, and focal glomerulo-

nephritis. Granulation tissue forms on the turbinates, causing nasal obstruction. Nasal intubation, even for the insertion of a nasopharyngeal airway, may be impossible. The organ most devastated by this disease is the kidney and the patient in irreversible renal failure may need dialysis. An anaesthetic for renal transplantation may be required in these cases.

Lethal midline granuloma

In this rare and fatal disease corticosteroids and radiation therapy occasionally impede the destructive process.

The role of the anaesthetist is important in the above diseases because establishment of a good airway requires careful advance planning, sometimes including pre-operative tracheostomy. Pulmonary involvement may present a formidable anaesthetic risk, especially if arterial oxygen desaturation exists. Regional anaesthesia and supplemental oxygen may have to be considered.

Sarcoidosis

This systemic multisystem granulomatous disease characterized by spontaneous and complete remissions in the early stages, and by a slowly progressive course if the disease persists, may affect any tissue or organ. Pulmonary involvement is remarkably common in sarcoidosis, 88% of the patients presenting with an abnormal chest radiograph. Three distinct areas of pulmonary lesions have been described: pleural, peribronchial and septal. Enlarged hilar lymph nodes may cause bronchial obstruction and distal atelectasis. In general, the total lung volume is usually diminished with pulmonary sarcoid infiltration, and the diffusing capacity tends to fall early. The upper airway may also be involved. The most common cardiovascular complications of sarcoidosis is right ventricular enlargement, but sarcoid lesions have occasionally been found in the valves and more often in the myocardium, sometimes resulting in arrhythmias or heart block.

Pulmonary and cardiac involvement are major concerns for the anaesthetist. It is important to define the status of pulmonary function before operation in patients with evidence of

pulmonary sarcoidosis. Pre-operative cardiac evaluation is also mandatory. In particular, evidence of pulmonary hypertension, right ventricular hypertrophy and any arrhythmias should be sought. Other associated abnormalities in sarcoidosis which may be of importance to the anaesthetist are as follows: (i) 20% of patients may be hypercalcaemic; (ii) 50% have abnormal liver function; (iii) 50-75% have hyperglobulinaemia; (iv) thickening of the septal and turbinate mucosae may lead to nasal obstruction; (v) laryngeal involvement is possible¹² (the lesions are typically granulomas or nodules involving the entire larynx or the supraglottic larynx alone, airway obstruction is possible, and a tracheostomy may become necessary in some acute cases); (vi) the recurrent laryngeal nerve is among the cranial nerves attacked by this disease, with resultant unilateral vocal cord paralysis; and (vii) the patient may have tracheal stenosis.¹³ Often patients with sarcoidosis have been on long-term treatment with corticosteroids and will need pre-operative coverage with bigger doses.

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News and Comment/Nuus en Kommentaar

Epidemiology of peptic ulceration

There has always been something unsatisfactory in our understanding of the causation of peptic ulceration. Although much is known about its pathophysiology, the exact aetiology is still obscure. A recent epidemiological study in Australia has shown striking regional differences in the incidence of peptic ulceration which may indicate that some unknown ulcerogenic factor is responsible (Hugh *et al.*, *Med J Aust* 1984; **141**: 81). This analysis was carried out by studying prescriptions for cimetidine issued through the Pharmaceutical Benefits Scheme, hospital admissions, and deaths in a population of 13 million Australians in 1981. The average incidence of peptic ulceration was 3,8/1 000 for duodenal ulcers and 0,7/1 000 for gastric ulcers.

Seventy thousand patients received initial treatment each year, two-thirds of them outside hospital. Patients with gastric ulcers were more likely than those with duodenal ulcers to be admitted to hospital or die. Striking differences were found in the incidence of peptic ulcer in the different states, the inhabitants of New South Wales being four times more likely to develop the condition than those in neighbouring Victoria. There seemed to be no reason for differences in prescribing habits between the two states (which would invalidate these results), and an examination of possible differences in diagnosis and treatment between the different states failed to reveal any obvious reasons for the differing incidence of the disease. The authors concluded that peptic ulcer results from a combination of environmental factors and inherent susceptibility, and that further investigations are needed.