

The implications and management of acute odontogenic infection in association with Down and Eisenmenger syndromes and schizophrenia in a rural setting

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Abstract

Background: This report describes the case management of a 32 year old special needs patient with life-threatening odontogenic infection. The combination of schizophrenia, Down and Eisenmenger syndromes presented significant challenges to managing his oral health, particularly within the rural context. In this case, dental treatment was limited to a full dental clearance during a high risk general anaesthesia session.

Method: A comprehensive work-up prior to general anaesthesia was an essential aspect of care. This included a full medical history and examination, communication between medical specialists, the dentist and family consultation. The anaesthetic procedure was undertaken using a careful regimen of drugs and monitoring to minimize the impact on his cardiovascular system. Techniques to minimize bleeding from extraction sites were also important.

Results: Three weeks postsurgically the patient was reviewed and his family reported that he was interacting positively with them after years of surliness and conflict. This was attributed to a managed psychotropic medication regimen and improved dental condition, which has led to a sustained improvement in quality of life.

Conclusions: The management of acute odontogenic infection for special needs patients in the rural setting requires a local interdisciplinary team approach, careful consideration of related pathophysiology and its potential impact on general anaesthesia, and close consultation with family and carers.

Key words: Special needs dentistry, odontogenic infection, Eisenmenger syndrome, interdisciplinary approach, rural setting.

Abbreviations and acronyms: AD = Alzheimer's disease; TOE = transoesophageal echocardiography.

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INTRODUCTION

This case study reports on the care of a patient with the complex association of Down and Eisenmenger syndromes and schizophrenia whose health was significantly compromised by acute odontogenic infection. His medical history presented significant challenges to managing his oral health, particularly within the rural context, which offers fewer health services, providers and referral options.¹⁻⁴ The case highlights the potential seriousness of odontogenic infection and importance of an interdisciplinary approach to special needs dentistry based on good communication between local dental and medical practitioners, family and carers, as an essential aspect of practice.

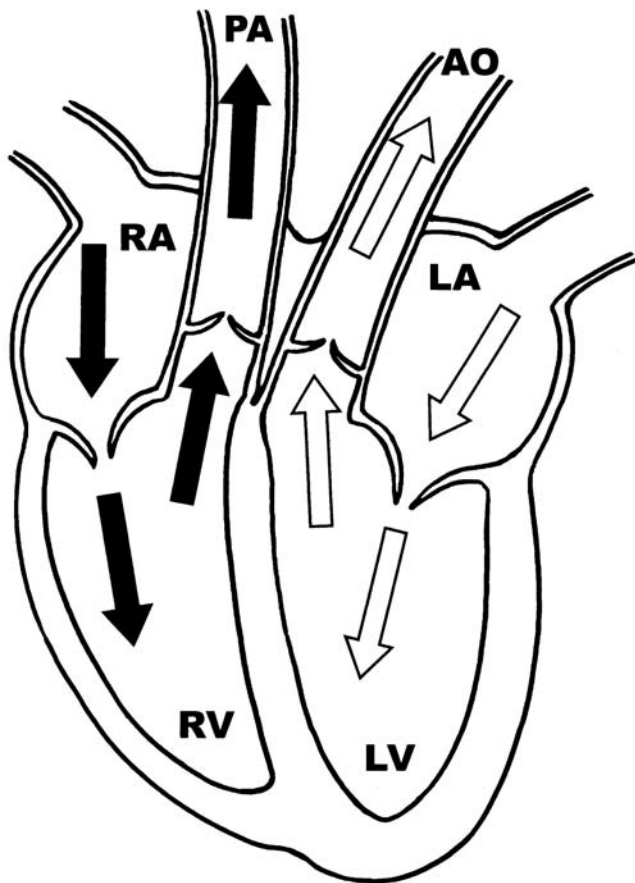
About 40 to 50 per cent of Down syndrome patients have significant congenital heart defects, such as mitral valve prolapse and atrial or ventricular septal defects.^{5,6} Due to the high relative incidence of large septal defects, Eisenmenger syndrome has been found to be most often associated with the Down syndrome population.⁷ The condition arises when blood flow through the heart defect increasingly elevates pulmonary vascular resistance until there is shunting of the deoxygenated blood from the right side of the heart to the systemic circulation. The progression to Eisenmenger syndrome in the presence of ventricular septal defect is illustrated in the series of Figs 1-3.

The pulmonary hypertension associated with Eisenmenger syndrome precludes correction, as the elevated pulmonary vascular resistance persists or worsens after surgical closure of the septal defect.⁸ Over time, the body compensates for the low oxygen content of circulating blood by increasing the red cell count. The resultant raised blood viscosity increases the risk of thromboembolic complications. Once Eisenmenger syndrome is established, there is progressive fatigue, dyspnoea and cyanosis. Overall, life expectancy is reduced with most patients dying from ventricular arrhythmia, thromboembolism or complications of non-cardiac surgery such as "dental, ophthalmic, or simple outpatient surgery".⁸

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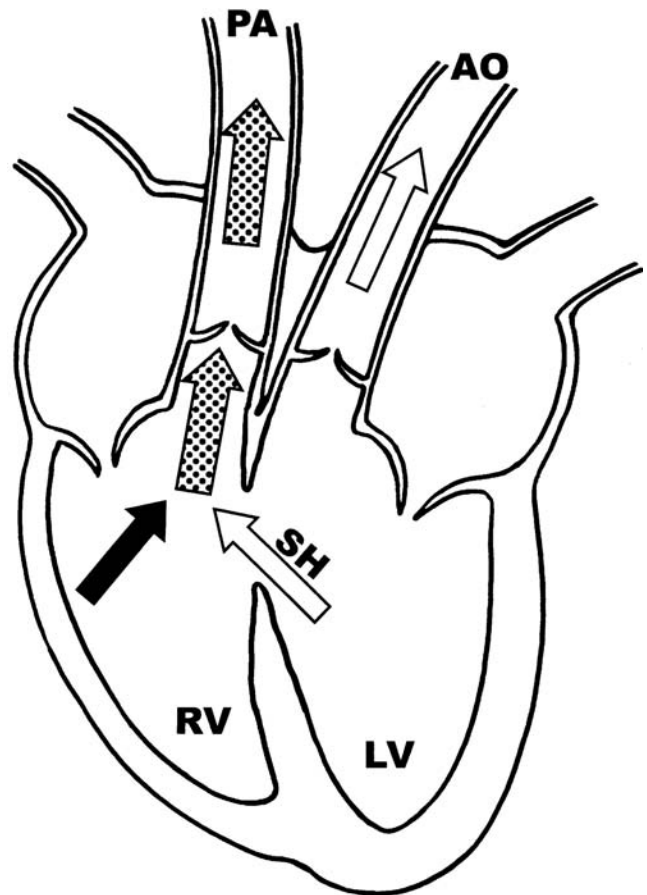
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BLOOD FLOW Deoxygenated 
Oxygenated 

Fig 1. Flow of blood through the normal heart. Deoxygenated blood is collected in right atrium (RA) and passes to the right ventricle (RV) to be pumped to the lungs via the pulmonary artery (PA). Oxygenated blood returns from the lungs and is collected in left atrium (LA) and then leaves the left ventricle (LV) to the body via the aorta (AO).

Down syndrome patients with Eisenmenger syndrome require special consideration in the delivery of their dental care.⁵⁻⁸ Vongpatanasin *et al.* conducted a literature review on Eisenmenger syndrome in adults, which noted that non-cardiac surgery should be avoided where possible since the administration of general anaesthesia in these cases may be associated with a peri-operative mortality rate of up to 20 per cent.^{8,9} Many of the agents used for induction and maintenance of general anaesthesia depress myocardial function and reduce systemic vascular resistance, with a resultant increase in the magnitude of the right to left shunting and cyanosis.⁹⁻¹¹ This situation may be difficult to reverse and can lead to cardiac arrest. Similarly, outpatient conscious sedation carries the risk of increasing hypoxia that may be difficult to reverse.⁹ Other precautions include minimizing bleeding and prompt fluid replacement for any blood loss that does occur. Antibiotic prophylaxis is necessary for the prevention of bacterial endocarditis,¹² as is early postoperative mobilization to prevent thromboembolism.






BLOOD FLOW Deoxygenated 
Oxygenated 
Mixed 

Fig 2. Flow of blood through the heart in the presence of a ventricular septal defect. Oxygenated blood is shunted (SH) through a hole in the wall separating the right and left ventricles from the more powerful, higher pressure left ventricle (LV) to mix with deoxygenated blood in the right ventricle (RV). This results in an increased blood flow through pulmonary artery (PA) and distal pulmonary vessels.

CASE REPORT

Initial dental consultation and assessment

A 32 year old male with Down syndrome and large congenital ventricular septal defect who lived in his familial environment presented to a public dental clinic in rural Tasmania for a dental examination. He had moderate intellectual impairment, minimal speech and a recent history of becoming progressively resistant to social interaction, agitated and disruptive. At examination, the patient repeatedly rocked his body and gestured to his mouth. Although it was not possible to conduct a comprehensive oral examination, there was evidence of extensive dental caries, generalized chronic soft tissue infection and extremely poor oral hygiene. The patient had attended the same public dental clinic at two yearly intervals since undergoing general anaesthesia, without complication, to remove four teeth and restore 16 teeth six years previously.

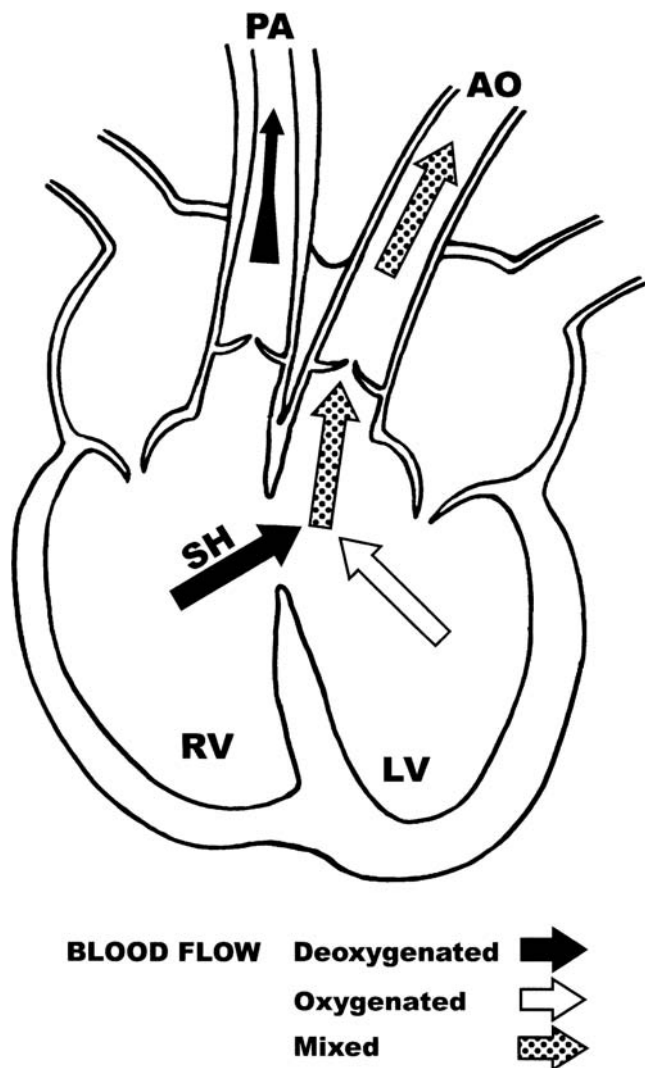


Fig 3. Flow of blood through the heart in the presence of ventricular septal defect and the development of Eisenmenger Syndrome. Over time, the increased pulmonary blood flow damages the pulmonary vessels which become narrowed. As a result of this narrowing, there is an increase in pressure in the pulmonary artery (PA) and right ventricle (RV). This pressure eventually reverses the direction of blood flow through the hole in the ventricular wall. There is now a shunt (SH) of deoxygenated blood from right ventricle (RV) into left ventricle (LV), where it dilutes the oxygenated blood pumped out the aorta (AO) to the rest of the body.

With the deterioration of his oral health it was felt treatment under general anaesthesia was indicated and specialist medical advice was sought on his present fitness to undergo this course of action.

Medical assessment

The patient was referred to a cardiologist who indicated that the patient's cardiac condition had deteriorated. An echocardiogram suggested his pulmonary circulation pressure was near his systemic pressures and there was bidirectional flow through the ventricular defect, i.e., early Eisenmenger syndrome. A warning was stated that situations that could bring about vasodilation, such as during general anaesthesia, could cause a marked right to left shunt, severe hypoxia with the possibility of triggering a lethal arrhythmia

(DR McTaggart, written communication, March 2005). Anaesthetic opinion from the tertiary hospital in Hobart confirmed the extremely high anaesthetic risk. With this knowledge, his family was understandably hesitant to proceed with the general anaesthesia.

Psychiatric assessment

The patient's psychiatrist had recently diagnosed him with schizophrenia and commenced Olanzapine wafers (5mg daily), which is an "atypical" antipsychotic drug (P Wurth, written communication, April 2005). Family members stated that he was frequently aggressive and strongly resisted attempts to provide assistance with home oral health care. His refusal to drink water and regular consumption of high levels of cariogenic drinks compounded the problems associated with managing his oral health. He was known to be fearful of medical procedures and his intellectual impairment and schizophrenia made procedures under local anaesthesia or sedation impractical and venepuncture problematic.

Subsequent presentation at a regional hospital

During the period of medical specialist consultation, the subsequent development of a large submandibular, acute dental abscess significantly increased the risk of bacterial endocarditis and the potential for airway obstruction. Pharmacologic management of this serious infection in the first instance was less than ideal.¹³ Oral Amoxicillin (500mg three times a day) for 10 days was prescribed by hospital staff. Although an intravenous Amoxicillin and Metronidazole regimen was considered, the hospital staff chose a longer course of treatment with oral Amoxicillin due to the patient's profound reluctance to intravenous cannulation or intramuscular injection and a previously noted intolerance to oral Metronidazole. The spread of infection was monitored and contained, with the family and dental practitioner remaining in daily contact until a subsequent anaesthetic assessment took place (DR Butler, written communication, July 2005).

Final anaesthetic assessment

Assessment by a specialist anaesthetist at a regional teaching hospital found the patient had a fair exercise tolerance, could walk slowly around without shortness of breath and did not suffer cyanotic or fainting episodes (CR Chilvers, written communication, July 2005). On examination, the patient weighed 59kg, his pulse was 72, blood pressure 112/82mmHg, and oxygen saturation on air was 98 per cent. Blood testing found haemoglobin to be high at 176g/l (normal 140–180) and mean corpuscular volume (MCV) to be raised at 99fl (normal 80–96), indicative of an increased number of immature red cells. Although not above maximum levels, taking into account his medical history and history of poor eating, this data indicated the presence of polycythemia and confirmed the severity of his cardiac condition. An opinion was sought about transferring him to a cardiac anaesthetic

unit at a major interstate hospital. This was not progressed as it was felt that this would result in only marginal improvement in outcome and greater disruption and distress for the patient and his family.

An anaesthetic plan was formulated to include a generous oral premedication and family accompaniment at induction.¹⁴ Other important considerations included avoidance of prolonged fasting, antibiotic prophylaxis, and care with intravenous injections to avoid systemic air embolism. Invasive cardiac monitoring with pulmonary artery catheter or transoesophageal echocardiography (TOE) was considered but evidence for efficacy is lacking.¹¹ In addition it was felt the use of this equipment would prolong the procedure unduly and in the case of TOE, obstruct access for dental procedures. Ultimately, it was planned to use only intra-arterial pressure monitoring and to use an anaesthetic regimen that minimized vasodilation and reduced right to left shunting. The family were advised that he would have around a 90 per cent chance of surviving such an anaesthetic.

Consent issues

It was evident to the dentist coordinating the patient's interdisciplinary assessments that the treatment of choice was limited to expeditious full clearance, including two unerupted third molars, under general anaesthesia. This decision was taken to avoid the risk of future general anaesthetic sessions since Eisenmenger syndrome patients' health deteriorates over time and the likelihood of reoccurrence of dental disease given the difficulties in maintaining his oral health. The patient's family was given an explanation that without operative treatment his symptoms would be ongoing and that death from recurrent odontogenic infection may be the ultimate result. They were

counselled that although the risks of general anaesthesia were high, they were not prohibitive. With the risk of oral infection removed, it was likely that improved quality of life would be sustainable during his 15 year life expectancy. The family agreed on this course of action.

General anaesthetic session

The anaesthetic preceded with premedication of Ketamine 400mg and Midazolam 20mg in 50ml of cola soft drink (the beverage most acceptable to the patient).¹⁴ Within 30 minutes, an intravenous line was able to be inserted, an oxygen mask applied and monitoring by pulse oximetry begun. During transfer to the operating theatre he required chin lift to relieve minor airway obstruction that had precipitated right to left shunting and resulted in a rapid decrease in oxygen saturation.

In theatre, an infusion of the vasoconstrictor Metaraminol was commenced for treatment of systemic arterial hypotension should this arise. Two litres of Hartmann's solution were given during surgery to maintain hydration. Intravenous Amoxicillin 1g was given prior to the procedure and afterwards in the recovery ward. Induction of anaesthesia was by a further Ketamine 50mg, Fentanyl 225µg, and the muscle relaxant Rocuronium 40mg given intravenously. A reinforced cuffed oral endotracheal tube was placed and anaesthesia maintained with oxygen and Desflurane delivered by gentle positive pressure ventilation. Pulse, blood pressure, oxygen saturation and other vital signs remained stable throughout the procedure.

Twenty-six teeth and two roots were extracted using elevators, luxators and forceps (Fig 4). Following



Fig 4. OPG.

consultation with the anaesthetist as to the patient's anaesthetic status, both lower unerupted third molars were removed surgically, with removal of buccal bone. Pre-emptive local block and infiltration anaesthesia with the administration of 6.6ml Citanest with 3% Octapression was utilized to additionally assist with haemorrhage control and avoid the use of adrenaline. Extraction wounds were closed using 3/0 Vicryl sutures. Eight Gelatamp sponges (with combined antimicrobial and thrombocyte aggregation capabilities) were used to pack the surgical sites (four each site). Paracetamol 1g as suppositories was also given to minimize postoperative pain.

On awaking from anaesthesia the patient was taken to the recovery ward where his oxygen saturation levels and blood pressure were monitored closely. Oxygen was administered for one hour but when removed, saturation levels dropped to 78 per cent. Although he was reluctant, oxygen was re-administered over the next two hours until saturations were maintained in the mid 90s on room air. He remained very drowsy and had some nausea and vomiting. There was "slight ooze" from the surgical extraction sites. The postsurgical "ooze" persisted and was monitored for five hours postoperatively but there were no ongoing complications. The patient was admitted overnight for observation but within 24 hours was well and alert enough to be discharged. He was placed on a soft food diet and no plans were made for dentures to be made for the immediate future.

Outcome

Three weeks postsurgically and at 12 month follow-up, his family members reported that he was interacting positively with them after years of surliness and conflict and there were strong indications of his improved quality of life. His psychiatrist attributed these improvements to his antipsychotic medication and improved dental condition (P Wurth, written communication, August 2005).

DISCUSSION

The majority of reported studies on the prevalence of dental caries and periodontal disease among Down syndrome populations have included relatively young populations and are based mainly on cross-sectional studies. However, the prevalence of dental caries among Down syndrome individuals is generally considered to be low compared to different control groups, with those individuals residing in institutions generally having a lower caries experience than those residing at home.¹⁵⁻¹⁸ The reasons for this observed lower caries rate are varied and may include low levels of *S. mutans* found in the saliva, genetic or environmental factors.^{16,17} The increased prevalence of periodontal disease among people with Down syndrome compared to age matched groups of both non-Down syndrome and other people with intellectual disability is well documented.^{5,7,15,19,20} The mechanism

for this is unknown, although impaired cell mediated immunity, disturbed serum immunoglobulins and a corresponding decrease in the number of T cells is thought to play a part.²⁰ Other contributory factors may include poor manual dexterity, poor oral hygiene, and hypotonicity of facial muscles, an open mouth posture and mouth breathing leading to reduced cleansing capacity of saliva.^{19,20} A number of studies have indicated that the levels of plaque and calculus seen in Down syndrome patients were not related to the severity of the periodontal condition.⁵ A number of recent studies have demonstrated that individualized preventive dental care, performed regularly using generally available methods and assistance with maintaining adequate oral hygiene are effective for suppressing the severity and progression of periodontal disease in Down syndrome patients.¹⁸⁻²¹ However, as is evident from this case study, the wide variance in the degree of intellectual impairment and the development of complex medical histories may present significant barriers to achieving and maintaining oral health.¹⁹

Severe odontogenic infection is potentially a life-threatening event and "management is primarily surgical with skilled anaesthetic airway management".¹³

Over the last 10 years, four patients in whom severe infection has resulted in airway obstruction have received care at the regional teaching hospital used in this case study. One case required weeks of treatment in intensive care for sepsis and one ultimately died from necrotizing fasciitis.

There have been considerable improvements in the historically low life expectancy of people with Down syndrome, with 80 per cent of adults living to 55 years of age or over.²² However, the development of Eisenmenger syndrome in association with ventricular septal defect will still lead to significant limitations in health and longevity. Interestingly, the most significant health risk for people with Down syndrome over 40 years of age has been identified as Alzheimer's disease (AD).²² After 35 years of age, virtually all individuals with Down syndrome have developed the neuropathologic characteristics of AD, although a smaller percentage undergo symptoms of cognitive decline.²² The general dental practitioner should be aware of the possible onset of clinical symptoms of AD, since adult patients may experience memory loss, be difficult to manage behaviourally in the clinical setting, and the level of home care may decline.

The optimal management of dental care for the special needs patients requires a well-coordinated interdisciplinary approach, which can only be successful with goodwill and good communication. In this case, decisions were made after consultations between the dentist, psychiatrist, cardiologist and anaesthetist.

CONCLUSIONS

Early prevention and oral environment management strategies to minimize the risk of potentially severe

odontogenic infections are highly desirable. However, for increasing numbers of people with Down syndrome living at home with elderly parents, effective home care and continuity of oral health care may be difficult to achieve. Adult Down syndrome patients may also present with increasingly complex medical histories. Although the implications of acute odontogenic infection were recognized in this patient, the potential seriousness of this condition may be underestimated. In this case, treatment options were determined by a local interdisciplinary clinical approach and were limited to full dental clearance under high risk general anaesthesia. Consent was a key issue and family/carers were involved in all decisions. The successful outcome was due to consideration being given to the pathophysiology of Eisenmenger syndrome, appropriate modifications of general anaesthesia technique, the management of his psychiatric condition and the permanent removal of the burden of dental infection.

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