



# Acute Rheumatic Fever and Rheumatic Heart Disease: The Implications on Oral Health

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## Abstract

Acute Rheumatic Fever (ARF) is an inflammatory disorder caused by an autoimmune response to group A streptococcus bacteria. Rheumatic Heart Disease (RHD), the condition caused by ARF, leads to long term damage of the heart valves. Although rare in developed countries, there are approximately 0.5 million new cases globally of ARF each year and around 300,000 of these patients will develop symptoms of RHD.

This report details a 7-year old girl with no previous cardiac history who developed ARF in September 2019 where she presented with coughing, fever, murmur and Group A streptococcus positive throat swab. She then rapidly developed RHD leading to moderate mitral valve regurgitation, mild mitral stenosis and mild aortic stenosis.

The patient was referred by the Cardiologist team to the Paediatric Dental department following diagnosis of RHD. On examination, the patient had caries in six primary teeth: 54, 55, 64, 65, 74, 84. Although the patient was ASA 4, the family and the wider team agree GA is the only option to facilitate the completion of the required dental treatment. The removal of potential sources of infection was a prerequisite to allowing the patient to proceed with her urgent cardiac surgery. Comprehensive dental care under general anaesthetic with support from 2 cardiac anaesthetists was undertaken to minimise the delay of urgent cardiac surgery.

Due to the increased risk of recurrent caries, the patient is kept on a three month recall within the department. She underwent an aortic valve repair and mitral valve replacement surgery in November 2019. She has recovered well since her operation and remains under regular review with her cardiologist.

Members of the dental team should be aware of the implications of ARF on child dental health. Many of these children will develop RHD. It is vitally important that these children remain caries free to prevent the devastating effects of rheumatic heart disease and infective endocarditis.



## Introduction

Rheumatic heart disease (RHD) is the condition caused by acute rheumatic fever. Acute rheumatic fever (ARF) itself is caused by the autoimmune response to Group A Streptococcus (GAS) infections [1]. This response leads to long term damage to the heart valves. There is currently no effective vaccine against ARF.

ARF is rare in developed countries. The introduction of antibiotics and improvements to living conditions and sanitation has led to the decline of ARF cases in developed countries. Overcrowding has been shown to increase the risk of GAS infections and ARF [2]. According to the World Health Organisation, there are at least 15.6 million people with RHD. There are approximately 0.5 million new cases of ACF each year and around 300,000 of these patients will acquire RHD. There are 233,000 deaths worldwide annually attributed to ACF or RHD [3].

The incidence of ARF in indigenous populations in Australia and New Zealand is 80-100 per 100,000. This can be as high as 245-351 per 100,000 in Aboriginal populations [1]. Around 60% of these patients will develop RHD [1]. The incidence of ARF in the non-indigenous population of New Zealand is 10 per 100,000 [1]. ARF most commonly occurs before adolescence and is rare in children under 5 years old and adults above the age of 35. The prevalence of RHD increases with age and peaks between 25-34 years. ARF and RHD are more common in females than in males [1].

Recurrent or severe acute rheumatic fever can cause damage to the cardiac valves. This is known as rheumatic heart disease and can increase the risk for further cardiac conditions such as infective endocarditis, stroke and congestive heart failure [4].

## Case report

### Diagnosis

Group A streptococcal (GAS) sore throat is the infection that precedes ARF. It should be differentiated from non-streptococcal pharyngitis and treated (see below). Patients will present clinically with fever, sore throat with pustules, petechiae on the palate and tenderness of the cervical lymph nodes [5].

The differential diagnoses for ARF should include diseases such as septic arthritis, Lyme disease, sickle cell anaemia, infective endocarditis, leukaemia and lymphoma [6].

Investigations include throat culture, rapid streptococcal antigen test, antistreptolysin O (ASO), erythrocyte sedimentation rate (ESR), C-reactive protein (CRP), full blood counts, chest X-ray and electrocardiogram (ECG). ECG will show transient prolongation of P-R interval [7].

ARF occurs 10 days to 6 weeks post initial streptococcal infection [7]. ARF is diagnosed if the patient has two major or one major and two minor criteria as described by Jones in 1944 [8]. This will be supported by history and evidence of previous GAS infection. Major criteria include: carditis, polyarthritis, chorea (involuntary muscle movement), subcutaneous nodules and erythema marginatum. Minor criteria include fever, polyarthralgia, polymorphonuclear leucocytosis and prolonged P-R interval on ECG.

### Treatment

Antibiotics are the treatment of choice for group A streptococcus infections. Antibiotic therapy should begin no more than

9 days after symptoms present [3]. Aspirin is the drug of choice for anti-inflammatory treatment of arthritis and carditis caused by ARF [5]. For moderate to severe carditis, steroid treatment (prednisolone) is recommended [5]. There is an improvement of the carditis over several years if there are no ARF recurrences [9]. Recurrence rate is 8-10% within 5 years with greater valvular damage with each recurrence [10].

In the long-term, benzathine penicillin G intramuscular injections are recommended to prevent recurrence. Doses are administered every 3-4 weeks for 10 years or until the age of 21 [4]. The purpose is to prevent colonisation of GAS and development of recurrent ARF. If there is no carditis, secondary prophylaxis can be discontinued after 5 years whereas in severe case secondary prophylaxis should be lifelong or up to the age of 40 [5]. This presents a significant burden on families and patients, especially in rural communities where access to healthcare is limited.

Surgery may be required to manage severe valvular damage from RHD. Active rheumatic carditis is an important predictor of valve failure and the need for repeat surgery [6]. Valve repair is usually the surgery of choice, but valve replacement may be required.

### Clinical presentation

A 7-year-old female (KR) was referred by her cardiologist to the Paediatric Dental department. She complained of occasional dental pain from the lower right quadrant. She did not require regular analgesics. The patient KR had been diagnosed with RHD one month prior to her dental appointment. As a result of RHD, she had developed moderate mitral valve regurgitation, mild mitral stenosis and mild aortic stenosis. She had been prescribed furosemide, spironolactone and penicillin V. She has no known allergies.

KR had no previous history of dental treatment other than preventative treatments by her general dental practitioner. She brushes twice daily with a manual toothbrush and 1450ppm fluoride toothpaste.

Extra-oral examination was normal and soft tissues were healthy. She was in the age-appropriate stage of the mixed dentition stage with first permanent molars, central incisors and lateral incisors fully erupted. There was clinically obvious caries affecting six primary teeth: 54, 55, 64, 65, 74, 84. Radiographic examination confirms caries in six primary teeth as well as periapical radiolucency present around 74 and 84.

The examination findings were discussed with the patient's family and extractions of the carious primary molars advised. Due to the patient's limited dental experience, the number of carious teeth and the urgency to complete treatment, the decision was made, in conjunction with the family and the cardiology and anaesthetic teams to complete the treatment under general anaesthetic. Although this patient was high risk for GA the removal of any potential source of infection was a prerequisite to allowing the patient to proceed with her urgent cardiac surgery. A comprehensive general anaesthetic treatment plan of examination under anaesthesia, extraction of carious primary molars and fissure sealants of the first permanent molars and non-carious primary molars was completed.

KR underwent aortic valve repair and mitral valve replacement in November 2019. Repair of the mitral valve was initially planned which was unsuccessful intraoperatively. The mitral

valve was instead replaced with a 25 mm mechanical prosthesis. The patient is now on warfarin anticoagulation therapy and Group A streptococcus prophylaxis with penicillin V. She has made a good recovery and is on a three month recall with her cardiologist.

The importance of oral health and caries prevention advice was provided and reinforced at an early stage. The patient is now on a three-month recall [11] within the Paediatric Dental Department for continued enhanced prevention [12].

### Conclusion

ARF and RHD are very rare in developed countries. Management of these diseases is complex and places a large burden on patients and their families. RHD may require valve surgery to repair or replace severe valve damage. Valve surgery places these patients at risk of infective endocarditis. Early dental assessment and treatment to ensure optimal dental health are vital to reducing the risk of infective endocarditis. Paediatric patients should be referred to specialist teams for assessment if possible, which allows for easier liaison with cardiology teams. Dentists should continue to be aware of the dental implications for patients suffering with RHD and the disease burden that entails.

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