Valvular heart disease is changing – a challenge for Africa

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The pattern of valvular heart disease is changing in Western populations [1]. There are implications for Africa as healthcare improves and people live longer.

Over the last half century in Western countries there has been a change in the incidence of valvular heart disease from a rheumatic cause to one of degeneration. Until the age of 64 years all **moderate to severe** valve disease affects less than 2%. In the group aged 64 – 75 years the proportion increases to 4 - 8% and after age 75 years it rises to 12 - 13%. **Mitral incompetence (regurgitation) and aortic stenosis contribute to the majority of cases. Mitral stenosis** is much more common in patients who have had rheumatic heart disease. As the population ages the healthcare burden of valvular heart disease will become greater.

Mitral incompetence is commoner than aortic stenosis. There are two main causes:

- The first, **primary,** arises from an abnormality of the valve.
- The second, **secondary**, follows an abnormality of the left ventricle.

In primary mitral incompetence the valve leaflets do not come together adequately at the time of systolic contraction of the left ventricle. The result is that some of the blood that should have been pumped out (i.e. a proportion of the stroke volume) is regurgitated into the left atrium. In severe cases this regurgitant volume may exceed 50% of the stroke volume. The left atrium then dilates. The left ventricle compensates by dilating to increase stroke volume following Starling's law of the heart. Initially no clinical symptoms may appear but eventually the left ventricle fails.

There are various forms of primary mitral incompetence:

- "Degenerative": this may lead to thickening or thinning of the valve leaflets. If the chordae tendinae lengthen or rupture then the leaflets prolapse into the left atrium.
- Bacterial endocarditis.
- Rheumatic cause is much less common than in developing countries.
- Congenital abnormalities of the valve.
- Drugs e.g. pergolide and carbergoline as used in Parkinson's disease [2].

The progression of primary mitral incompetence varies. About 50% of patients come to no harm. However at the opposite extreme about 20% develop heart failure. If the incompetence is severe when first diagnosed then the outlook is much worse. Also if the cause is a ruptured of chorda tendinae the prognosis is also poor.

Myocardial infarction or a cardiomyopathy leading to left ventricular dilatation or hypertrophy may cause a dilatation of the mitral ring and hence secondary mitral incompetence. The valve leaflets themselves remain normal but do not come together in systole. Ischaemic coronary artery disease (including myocardial infarction) is the commonest cause. So when the latter occurs it is on top of an already damaged left ventricle. About 12% of patients develop moderate to severe mitral incompetence a month after a myocardial infarction.

Aortic stenosis results from an impaired mobility of the valve cusps (normally three). The opening of the valve in left ventricular systole gradually reduces (normal 3 – 4 cm^2). Eventually the rising pressure gradient across the valve leads to extra work for the left ventricle. The ventricle compensates by increasing the muscular wall thickness (hypertrophy). Exertional breathlessness indicates failure of the left ventricle.

There are three main causes of aortic stenosis in Western countries:

- A bicuspid aortic valve is a congenital abnormality and is more prone to calcification.
- A normal tricuspid valve may become calcified.
- Rheumatic heart disease but this is now rare.

A number of factors predispose to thickening of the aortic valve (before stenosis occurs this is called sclerosis):

- Increasing age.
- Males more than females.
- Hypertension.
- Smoking.

- Diabetes mellitus.
- Hyperlipidaemia.

About a quarter of people over aged 65 years have a degree of aortic sclerosis. Almost 10% of these develop aortic stenosis during the next five years.

Aortic incompetence (regurgitation) occurs if the valve cusps do not come together closely in left ventricular diastole. The consequence is that some of the stroke volume flows back into the left ventricle in diastole. In severe cases this may reach 50% of the stroke volume putting extra strain on the left ventricle which then gradually dilates and the wall hypertrophies. For a time this creates a compensatory situation but eventually the left ventricle fails.

The underlying causes of aortic incompetence include:

- Abnormality of the valve itself e.g. congenital bicuspid structure, bacterial endocarditis.
- Aortic root abnormality especially if this is accompanied by a dilated root as in e.g. syphilis or Marfans syndrome.

Mitral stenosis results from a thickening and fusion of the valve leaflets. The opening of the valve (normal 4cm^2) is reduced during left ventricular diastole (left atrial systole). The pressure in the left atrium rises and this chamber dilates. The lung vascular pressure rises (pulmonary hypertension). The patient becomes breathless on exercise. Wheezing may mimic bronchial asthma and hence suggest the wrong diagnosis. Finally in turn the right ventrical dilates and signs of right ventricular failure with pedal oedema and eventually ascites do occur.

The cause of mitral stenosis in Western populations is now much more likely to be a congenital abnormality whereas in a developing rheumatic fever remains the overwhelming cause.

The situation in Africa

A review of valvular heart disease (rheumatic and non-rheumatic) in Africa [3] shows that **the focus remains on rheumatic heart disease.**

In Western nations valvular heart disease affects the older patient and is slow in onset. In Africa it is the young who tend to be affected and the disease progresses much more rapidly.

The Lancefield Group A streptococcus is the precipitating cause of acute rheumatic fever. In Western countries it occurs in less than 1 in 100,000 of the population whereas in Africa (e.g. Sudan) it is as common as 1 in 1,000. The reasons for this difference probably include:

- Household overcrowding, especially with increasing urbanisation, leading to closer person-to-person contact.
- Patterns and / effectiveness of antibiotic use.
- Virulence of the infecting streptococcus.

A review of some of the incidences of chronic rheumatic heart disease in school children is striking:

- Nairobi (Kenya): 2.7 in 1,000
- Kinshasa (DRC): 14.3 in 1,000
- Soweto (South Africa) (1975): 6.9 – 20 in 1,000
- Kampala (Uganda) (2012): 14.7 in 1,000 [4]

What happens to the heart in acute rheumatic fever

Echocardiographic studies indicate that severe mitral incompetence is the result of annular (the ring structure at the base of the valve) dilatation and elongation of the chordae tendinae leading to prolapse of the valve leaflet back in the direction of the left atrium. Sometimes the systolic murmur of mitral incompetence cannot be heard with the stethoscope and it is then that echocardiography is so useful.

Initially the valve leaflets are normal but later may become fibrosed and rigid adding stenosis to the incompetence.

This is what happens clinically in **acute rheumatic fever:**

- Streptococcal infection: usually a sore throat (pharyngitis).
- Up to three weeks later: fever and large joint pains and swelling.
- Fast heart rate (tachycardia) which seems greater than would be expected with the degree of fever.
- Systolic and diastolic mitral murmurs vary from time to time and frequent auscultation is mandatory.
- If pericarditis develops then a pericardial friction rub may be heard.
- Signs of heart failure suggest heart muscle damage.

However it must be appreciated that only about a quarter of patients who are found to have rheumatic valvular disease later in life give a clear history of rheumatic fever.

It is worth mentioning the importance of **rheumatic heart disease in association with pregnancy.** Where there is a significant rate of teenage pregnancy and a high

incidence of rheumatic heart disease the coincidence of each condition in the same patient is potentially serious. But which patients are at greatest risk? Mitral stenosis is the most common lesion, it is the most serious lesion and may be discovered for the first time in the antenatal clinic. In practice there are three main situations that may face the clinician and obstetrician:

- Known mitral stenosis and wishes to become pregnant.
- Mitral stenosis that is well compensated in a patient already pregnant.
- Severe mitral stenosis, a critical cardiac state (i.e. poor haemodynamic condition) and in advanced pregnancy.

The physiologically increased cardiac rate during pregnancy with its reduced diastolic filling time of the left ventricle poses the main danger for the pregnant woman with mitral stenosis. Wherever possible such patients should be referred urgently for a specialist opinion. The options include the use of diuretics, a beta-blocker, mitral valvuloplasty and valve replacement. However the surgical options are likely not to be available.

Non-rheumatic valvular heart disease in Africa

All of the non-rheumatic heart conditions that occur in Western populations may, of course, appear in African countries but there are a number of differences.

Bacterial endocarditis

The high prevalence of rheumatic valvular heart disease in Africa means that there are large numbers of patients at risk of bacterial endocarditis. This places a great burden on limited healthcare resources. Clearly efforts must be directed at the prevention of rheumatic valvular disease by prevention of the original rheumatic process caused by the streptococcus: namely the effective treatment of streptococcal infections and improved social conditions where such infections proliferate.

In addition the sources of infection for the endocarditis must be managed. For example, by adequate dental care and **appropriate** antibiotic use especially for streptococcal throat infections. In the UK, National Institute for Health Care and Excellence guidelines [4] no longer advise prophylaxis. There is no clinical evidence that prophylactic antibiotics reduce the incidence of infective endocarditis. The disadvantages (e.g. allergic reactions and increased bacterial resistance) may be greater than the benefits. But does this also apply in the same way to African circumstances [6,7]? Antibiotic-related side effects do occur but are uncommon after a single dose of antibiotics as for prophylaxis [8,9]. The American Heart Association [9] considers it safe to give a single dose of a broad spectrum antibiotic (e.g. amoxicillin) if there is no history of hypersensitivity. The controversy remains (see Author's note below).

Secondary prevention of rheumatic heart disease

This depends on adequate public health arrangements. Ideally benzathine penicillin 1.2 mega units once per month (or sulphadiazine 500mg twice daily) to those aged over 10 years and once daily to those under aged 10 years. The duration of this prophylaxis depends on how severe the original disease was and how long since the attack [10]. Usually the duration is taken as five years or until the patient is 18 years old.

Submitral Aneurysm

This condition is thought to be congenital in origin arising from a weakness in the mitral annulus. Most reports of this left ventricular aneurysm come from sub-Saharan Africa. It causes severe mitral incompetence because of undermining of the valve leaflets and papillary muscle. Sometimes the aneurysm ruptures into the left atrium. Cardiac dysrhythmias are common and myocardial ischaemia may be caused by compression of the left circumflex coronary artery. The only satisfactory treatment is surgical.

Finally, a quote directly from the publication by Essop and Nkomo [3]:

"Africa faces many difficulties, and the challenge of preventing and treating the scourge of rheumatic heart disease is enormous. If we are to rise to this challenge, we need to establish the scope and magnitude of the problem with large, properly conducted epidemiological trials, and health authorities must be urgently convinced of the need to institute efficient and readily accessible programs for the primary and secondary prevention of rheumatic heart disease."

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Author's note: There clearly is controversy about the use of antibiotics to prevent bacterial endocarditis in Africa. This needs further research. The clinical experience of colleagues in South Sudan reported to this journal would be most valuable: Is bacterial endocarditis often seen or recognised? Does it appear to follow interventions such as dental extractions?

STARLING, Ernest Henry (1866 – 1927) was a British physiologist studying at Guy's Hospital in London and later became Professor of Physiology at University College (London). He described what has become known as "Starling's law of the heart": the force of contraction of the heart muscle is proportional to its initial length. This means that, in for example mitral incompetence, as the left ventricle dilates and the muscle wall stretches the power (force) of contraction after each cycle of stretch increases. However eventually this "law" fails and so does the left ventricle. Another way of looking at this is to take an elastic band: stretch it to say 10 cms and release it and measure the distance it is projected. Then repeat the exercise but stretch the band to 20 cms before release: it should be projected approximately twice the distance. However if you then gradually increase the initial stretch further there will come a time when the band breaks. If you try this experiment be very careful not to flick the band into your eyes or someone elses!!!