Severe valvular and congenital heart diseases in adults

June 2008
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**Updating of doctors’ guides and lists for long-term conditions**

The long-term conditions guides produced by HAS are revised every three years.

This document may be downloaded from www.has-sante.fr
# List of abbreviations

<table>
<thead>
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<th>Abbreviation</th>
<th>Description</th>
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<tbody>
<tr>
<td>AC</td>
<td>anticoagulants</td>
</tr>
<tr>
<td>ACC</td>
<td>American College of Cardiology</td>
</tr>
<tr>
<td>ACEi</td>
<td>Angiotensin-converting enzyme inhibitor</td>
</tr>
<tr>
<td>ACS</td>
<td>Acute coronary syndrome</td>
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<tr>
<td>AF</td>
<td>Atrial fibrillation</td>
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<tr>
<td>Afssaps</td>
<td>French Health Products Safety Agency</td>
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<tr>
<td>AHA</td>
<td>American Heart Association</td>
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<tr>
<td>AR</td>
<td>Aortic regurgitation</td>
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<tr>
<td>ALD</td>
<td>Long-term condition (affection de longue durée)</td>
</tr>
<tr>
<td>ANAES</td>
<td>French National Agency for Accreditation and Evaluation in Healthcare</td>
</tr>
<tr>
<td>AS</td>
<td>Aortic stenosis</td>
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<tr>
<td>ASD</td>
<td>Atrial septal defect</td>
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<tr>
<td>AVB</td>
<td>Atrioventricular block</td>
</tr>
<tr>
<td>AVSD</td>
<td>Atrioventricular septal defect</td>
</tr>
<tr>
<td>BMI</td>
<td>Body mass index</td>
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<tr>
<td>BP</td>
<td>Blood pressure</td>
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<tr>
<td>CC</td>
<td>Consensus conference</td>
</tr>
<tr>
<td>CPG</td>
<td>Clinical practice guideline</td>
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<tr>
<td>CV</td>
<td>Cardiovascular</td>
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</tbody>
</table>
CVA  cerebrovascular accident
CV-RF  cardiovascular risk factors
ECG  electrocardiogram
EF  ejection fraction
ESC  European Society of Cardiology
HAS  Haute Autorité de Santé
HF  heart failure
HT  hypertension
INR  international normalized ratio
IUD  intra-uterine device
LA  left atrium
LMWH  low molecular weight heparin
LV  left ventricle
LVF  left ventricular failure
MA  mitral area
MR  mitral regurgitation
MS  mitral stenosis
MVP  mitral valve prolapse
NFH  non-fractionated heparin
PAH  pulmonary arterial hypertension
PAP  pulmonary arterial pressure
PG  polyglobulia
PR  pulmonary regurgitation
PMC percutaneous mitral commissurotomy
PS  pulmonary stenosis
RF  rheumatic fever
RV  right ventricle
SBP systolic blood pressure
SR  sinus rhythm
TR  tricuspid regurgitation
TIA transient ischaemic attack
TOE transoesophageal echocardiography
TTE transthoracic echocardiography
VKA vitamin K antagonists
VSD ventricular septal defect
Introduction

The aim of this guide for medical practitioners is to describe the optimal theoretical care and the clinical pathway for a patient entering the ALD [Long term condition] scheme, severe valvular heart disease.

Valvular heart diseases are very diverse and require different medical management. They range from severe congenital heart diseases which needed surgery in early childhood to those encountered among elderly. To provide a common overview in the same guide is definitely very simplistic.

This guide is intended to be a pragmatic reference tool for general practitioner, regarding the initial evaluation of patients with a severe valvular heart disease, its therapeutic management and follow-up. The contents of the guide have been discussed and validated by a multidisciplinary working group. It is a practical guide to available recommendations for clinical practice (RPC) or consensus conferences (CC), supplemented by expert opinions when data are lacking. Expert opinion is needed in fields such as patient follow-up, where the clinical management is based on consensus among professionals rather than on comparative data from clinical trials.

However, a guide for doctors on long-term conditions cannot cover all comorbidities and hospital care protocols. It cannot provide an exhaustive list of all possible management procedures or discharge doctors from their own individual responsibility towards their patients.

The prevalence of valvular heart disease in Western countries remains high, at around 2% and increases with ageing (prevalence of 10-15% among patients over 75).

An ageing population is resulting in the occurrence of dystrophic valvular diseases (mitral insufficiency, MI) and degenerative conditions (aortic stenosis, AS), while the frequency of rheumatism-related valvular diseases is gradually decreasing. Comorbidities must definitely be taken into account. Furthermore, in view of the advances made in paediatric heart surgery, children with complex congenital heart diseases are now reaching adulthood.

Valvular heart diseases are the third main cause of heart failure (HF). They induce also cardiac rhythm disorders, in particular in case of mitral stenosis (MS) and congenital heart diseases.

Infectious endocarditis is a serious complication, which seems to have a stable incidence of around 25-30 cases per million inhabitants, i.e. 1,500 cases a year in France. Prosthetic valve endocarditis is particularly serious. It affects 3-6% of patients, with the mortality rate varying between 20% and 40%.
The main sources used for drawing up this guide are the *Guidelines on the Management of Valvular Heart Disease* (European Society of Cardiology, 2007), the guidelines on the management of acquired valvular heart disease and prosthetic valve malfunctions (French Society of Cardiology, 2005) and the *Guidelines for the Management of Patients with Valvular Heart Disease* (American College of Cardiology/American Heart Association, updated in 2006). Their method of drafting recommendations differs from that of HAS (French National Health Authority).

The definitions used here for **serious valvular and congenital heart disease** are:

- Valvular heart diseases (stenosis or regurgitation) with:
  - severe valvular lesions (severe stenosis or regurgitation), or
  - less severe valvular lesions with:
    - symptoms of HF or ischaemic heart disease
    - in the absence of symptoms, objective proof of cardiac dysfunction at rest at echocardiography: reduced ejection fraction (EF), pulmonary arterial hypertension (PAH) or pronounced ventricular enlargement.

- Patients who have undergone heart surgery (prosthetic valves, conduits etc.).

- Complex congenital heart diseases operated on during childhood (tetralogy of Fallot, atrioventricular septal defect, coarctation of aorta, severe pulmonary insufficiency (PI) and pulmonary stenosis (PS)). They require a follow-up by a team experienced in congenital heart diseases.

- Highly complex congenital heart diseases operated on during childhood (transposition of the major vessels, truncus arteriosus, pulmonary or mitral atresia, univentricular heart disease, heart disease with PAH etc.). They require a regular follow-up by a team experienced in congenital heart disease.

HAS has produced other guides about heart diseases:
- *Atrial fibrillation* guide
- *Chronic symptomatic systolic heart failure* guide
- *Chronic symptomatic heart failure with preserved systolic function* guide

It has also produced guides about complex congenital heart diseases:
- Tetralogy of Fallot
- Transposition of the major vessels
- Truncus arteriosus

These documents can be consulted on the HAS website.
1. Initial assessment

The diagnosis of severe valvular or congenital heart disease is based on clinical examination and echocardiography.

1.1 Objectives
- Investigate the presence of symptoms
- Determine the severity of the valvular lesions
- Assess the prognosis
- Identify other related diseases
- Indicate whether intervention or follow-up is required
- Offer advice to patients and their families (prevention of infectious endocarditis etc.)

1.2 Professionals involved

The initial management of patients with valvular heart disease is ensured by the primary care doctor and/or cardiologist.

The management of operated complex congenital heart diseases requires follow-up at a congenital cardiological centre, with the frequency determined by the degree of complexity and the quality of the repair.

Where valve surgery is indicated, this involves a cardiac surgeon, anaesthetist and/or an interventional cardiologist.

In case of severe comorbidities, pregnancy or any discussion of non-cardiac surgery, a multidisciplinary consultation may be required.

1.3 Diagnosis

► Clinical examination

Symptoms, both current or in the patient's history and their progression are a key element in decision making. They must be quantified at rest or during exercise.
- Dyspnoea (NYHA class¹)
- Fainting
- Angina
- Rhythm disorders
- Signs of HF
- Thromboembolism

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1. New York Heart Association (NYHA) Classification:
   I. Asymptomatic during ordinary physical activity
   II. Limitation of physical activity for exertions in ordinary life
   III. Marked limitation of activity, with symptoms appearing in the case of moderate exertion
   IV. Permanent symptoms, even at rest
• Signs of infectious endocarditis (fever)
• Cyanosis (in the case of congenital heart disease)

Diagnosis is based on clinical evaluation, and a murmur can indicate the severity of the valvular disease. It is the main method for screening for valvular heart disease in asymptomatic patients. It investigates history and assesses comorbidities:

• Known history of heart disease
• Cardiovascular risk factors (CV-RF): smoking, HT, diabetes, dyslipidaemia, heredity, overweight
• Assessment of comorbidities
• Personal and family history

Paraclinical tests

• A transthoracic Doppler echocardiography is the key examination. It confirms the diagnosis, quantifies the severity of the valve lesion (gradient, valve area, size of leak) and assesses left ventricular function.

<table>
<thead>
<tr>
<th>Severity of valvular heart disease (defined by the echocardiography)</th>
</tr>
</thead>
<tbody>
<tr>
<td>AS</td>
</tr>
<tr>
<td>Severe valvular heart disease*</td>
</tr>
</tbody>
</table>

*The severity can only be confirmed by comparing the clinical criteria and all the echocardiogram measurements; adapted according to the ACC/AHA, 2006.

The echocardiography also indicates:

• Morphology and function of the valve
• Size of the left ventricle (LV), parietal thickness
• Size of the left atrium (LA)
• Pulmonary arterial pressure (PAP)
• Right ventricle (RV), especially in the case of congenital heart disease

It is used to calculate the EF, transvalvular flow velocities.

• The ECG is part of the consultation with the cardiologist and helps to evaluate the impact of valvular heart disease.
• A chest X-ray may be useful if there are inaugural signs.
1.4 Complementary tests
Additional examinations may be necessary, according to the cardiologist’s opinion:

► Transoesophageal echocardiography
Transoesophageal echocardiography (TOE) is indicated if:
• Echocardiography is inconclusive, especially if it cannot determine the mechanism of the mitral insufficiency
• Endocarditis, thrombosis or a prosthetic valve dysfunction is suspected
• In complex congenital heart disease, to investigate the cardiac morphology

► Other tests
Certain additional investigations may subsequently be useful. They require referral to a cardiologist or specialist team:
• Exercise test
to unmask objective occurrence of symptoms:
  ‣ Level of physical activity, blood pressure profile contraindicated in the case of symptomatic severe AS
• Holter ECG
to detect arrhythmias, particularly in complex congenital heart diseases
• Stress echocardiography (pharmacological)
in the case of AS with LV dysfunction
• Radionuclide angiography
to assess LV function in aortic or mitral insufficiency, especially when the echocardiography at rest is inconclusive
• Coronary angiography
  ‣ in the case of angina and/or suspected ischaemic heart disease
  ‣ when surgery is planned
Coronary angiography is indicated in men over 40 and women over 50, or in case of associated cardiovascular risk factors and when surgical correction of the heart disease is planned.
• Cardiac catheterization
  ‣ Restricted to the cases where the echocardiography has not provided sufficient information
  ‣ to measure pulmonary pressure and resistance (congenital heart diseases)
• MRI or scan
  ‣ in complex congenital heart diseases and for assessing the thoracic aorta

► Blood tests
• If there is a mechanical prosthetic valve or a rhythm disorder, especially atrial fibrillation (AF):
- haemogram with platelets
- prothrombin time if anticoagulant treatment with VKA is prescribed

- If clinical sign:
  - blood count
  - serum iron
  - blood creatinine, proteinuria
  - lactate dehydrogenase (LDH)
  - liver function test (complex congenital heart diseases)

### 1.5 Risk of infectious endocarditis assessment

The following patients are at high risk:
- Patients with prosthetic valves (mechanical, homograft or bioprosthesis)
- Patients with a previous history of infectious endocarditis
- All unoperated patients with cyanogenic congenital heart diseases and operated patients with congenital heart diseases where conduits have been implanted

Patients at an intermediate risk include those with:
- Non-cyanogenic congenital heart diseases, apart from corrected atrial septal defect (ASD) and ventricular septal defect (VSD)
- Acquired valvular heart disease and a bicuspid aortic valve
- Obstructive cardiomyopathy
- Mitral valve prolapse with a mitral leak and/or valve thickening on the echocardiogram
- Prosthetic aortic conduits

General hygiene measures must always be applied (high and intermediate risk), especially buccodental and cutaneous.

In the case of invasive procedures, antibiotic prophylaxis is recommended in the high-risk group and optional in the intermediate-risk group, according to the patient’s characteristics (ageing patient or with comorbidities) and doctor’s choice.

### 1.6 Thromboembolic risk assessment

Thromboembolic risk is linked to both the patient and valvular lesions.

The risks related to the patients are:
- History of thromboembolism, atrial fibrillation (AF)
- LA > 50 mm, intra-atrial dense contrast, MS, EF < 35%, hypercoagulability

The factors related to valvular involvement are:
- Mitral valvular disease
Mechanical prostheses
Patients with MS in AF or with a prosthetic mechanical valve are at high risk of thromboembolism. Certain complex congenital heart diseases (cavo-pulmonary shunts and related conditions) are at high risk, as well as cyanogenic heart disease (polyglobulia, PG).

1.7 Specific case of pregnant women or women wishing to become pregnant

Congenital and valvular heart diseases are the most common heart disorders found in women of child-bearing potential. Some specific heart diseases strictly contraindicate pregnancy due to the high mortality rate for the mother. A woman with a valvular heart disease wishing to become pregnant must have a preliminary assessment including a cardiological examination before becoming pregnant, and a close cardiac monitoring throughout her pregnancy. The pregnancy must be managed jointly by the cardiologist and the obstetrician.

The risk of complications for mother and child during pregnancy is less if management of the valvular heart disease has taken place before the start of the pregnancy. If a valve replacement is indicated, a bioprosthesis is the preferred choice, after informed consent of the patient and her family.

The risk of congenital heart disease of the child is higher than in the general population if the mother herself has a congenital heart disease.
2. Therapeutic management

2.1 Objectives

- Reduce symptoms and improve quality of life
- Prevent infectious endocarditis
- Prevent thromboembolism and the risks of anticoagulant treatment
- Manage cardiovascular risk factors
- Reduce mortality

The global strategy (intervention or follow-up) must be discussed with the patient, taking into account the following factors:

- Type of valvular heart disease
- Type and severity of symptoms
- Underlying heart disease
- Presence of AF
- Patient’s age
- Associated diseases
- Short and long-term treatment goals
- Constraints and follow-up of anticoagulant treatment

2.2 Professionals involved

The medical management of patients with serious valvular or congenital heart disease is under the responsibility of the primary care physician and the cardiologist. Other health professionals may be involved, particularly a cardiac surgeon, interventional cardiologist or a cardiologist with experience of congenital heart conditions in adults. A biochemist is involved in the event of anticoagulant treatment.

The follow-up of complex congenital heart disease must be carried out by a team experienced in congenital heart disease.

Pregnancy in women with valvular or congenital heart disease must be monitored by a cardiologist, in close collaboration with an obstetrician.

The patient’s therapeutic education can be provided by:

- Individual healthcare professionals (doctors, biochemists, nurses, pharmacists, dentists etc.)
- Managed care with a training programme
- Specific coordinated follow-up if complex congenital diseases
2.3 Self management education

► Therapeutic education
Self management education must ensure proper understanding of the patients (and their families) of their medical condition. This includes information about:
- Heart disease, its symptoms and risks, clarifying the warning signs that must lead to a medical visit
- Caution to be taken for contraception and if a patient wishes to become pregnant
- Follow-up visits and examinations
- Prescribed medications, possible adverse events of treatment
- Medications to be discontinued and to be avoided
Any change in or worsening of symptoms must result in a visit to the doctor.

► Lifestyle adjustments
- Learning nutritional rules (reduced salt intake if the patient shows signs of HF) and technical measures (self-monitoring of weight, heart rate, BP)
- Encouraging regular physical exercise adapted to the patient’s clinical condition; it may be limited, especially in the case of AS and complex congenital heart disease
- Advice on daily living, sexual activity, travelling, especially by air.
Managing the risk factors is important in the case of associated coronary disease:
- Reduction of obesity, cessation of smoking, management of metabolic disorders.

► Prevention of infectious endocarditis
General measures to prevent infections from various entries are always necessary: buccodental and cutaneous hygiene, disinfection of wounds, antibiotics treatment of any focus of infection.
Antibiotic prophylaxis during certain invasive procedures is recommended for high-risk patients and optional for patients at intermediate risk (see Section 2.4 on treatment).
The buccodental status of patients at an intermediate or high risk must be assessed at least twice a year. Any procedure which involves breaking the mucous membranes or the skin is contraindicated (body piercing, tattooing etc.).
The patient must carry an infectious endocarditis prevention card, which doctors can obtain from the French Federation of Cardiology (www.fedecardio.com).
**Monitoring treatment with vitamin K antagonists (VKA)**

When initiating a treatment with VKA, each patient must receive a specific education, explaining the purpose of the treatment, its risks, the principles of INR and adjusting the doses, possible adjustments to their lifestyle (professional and sporting activities), their diet (in particular with regard to foods rich in vitamin K), the main drug interactions and the risks of self-medication, the main signs of overdosage and underdosage, as well as explaining what to do in the event of an accident.

This education is initiated by the doctor or the unit indicating anticoagulant treatment. Depending on the patient’s abilities, the possibility of self-monitoring and self-management of the INR may be discussed.

The following points should be emphasised:
- Instructions about monitoring anticoagulant treatment (concept of target INR);
- Systematic reporting of oral anticoagulant treatment to all carers
- Patients must keep an up-to-date diary for monitoring their anticoagulant treatment, which doctors can obtain from the French Federation of Cardiology ([www.fedecardio.com](http://www.fedecardio.com)).

### 2.4 Pharmacological treatments

*ALD guides refer to drug classes without listing all the drugs indicated in the disease. Each drug is to be used only within the framework of its Marketing Authorisation. If for a specific reason this is not the case, and more generally, whenever a drug is prescribed in circumstances other than those given in the Marketing Authorisation, this is the sole responsibility of the prescribing physician, who must specifically inform the patient of this.*

**Antithrombotic treatment**

Anticoagulant treatment is essential in case of AF and in case of mechanical prosthetic valve:
- Lifetime treatment in the case of mechanical prosthesis
- Lifetime treatment in the case of a bioprosthesis and if there are risk factors linked to the patient, especially AF
- 3-month treatment post-surgery, in the case of a bioprosthesis or valve repair, with a target INR of 2.5 (range between 2 and 3)

Treatment with aspirin (75 to 100 mg per day) may be suggested as an alternative to VKAs, in the case of a bioprosthesis or valve repair, for 3 months after the operation (off-label).

When the patient has a mechanical prosthetic valve, anticoagulant treatment must be perfectly titrated, with a target INR that varies according to the type of prosthesis, its location and the thromboembolic risk factors of the patient.
The target INR varies between 2.5 (range 2 to 3) and 4 (range 3.5 to 4.5), depending on the circumstances (Appendix 2). As an example, if a patient with AF without a prosthetic valve and with valvular heart disease, the target INR is 2.5 (range 2 to 3).

The aspirin (75 to 100 mg per day) may be added, especially in the case of coronary disease or thromboembolism being treated with VKA.

The bleeding risk increases very quickly when the INR is > 4.5. An INR > 6 requires immediate medical management.

If the VKA treatment may need to be interrupted or switched to heparin, for diagnostic or therapeutic procedures, the specialist in charge of the planned procedures should seek for the advice of the cardiologist (cf. II.6).

► **Prophylactic treatment for infectious endocarditis**

The antibiotic prophylaxis depends on the procedure, the level of risk of the heart disease and the patient's characteristics.

The standard treatment in the case of dental, respiratory or oesophageal intervention includes: amoxicillin (which can be injected if unable to be administered orally; intramuscular injection is contraindicated in patients receiving anticoagulant treatment). For patients allergic to penicillin: pristinamycin or clindamycin.

<table>
<thead>
<tr>
<th>Condition</th>
<th>Product</th>
<th>Dosing</th>
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</thead>
<tbody>
<tr>
<td>No allergy to beta-lactam antibiotics</td>
<td>Amoxicillin</td>
<td>Single dose administered during the hour prior to the procedure</td>
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<tr>
<td></td>
<td></td>
<td>3 g <em>per os</em> (2 g if weight &lt; 60 kg)</td>
</tr>
<tr>
<td>Allergy to beta-lactam antibiotics</td>
<td>Pristinamycin</td>
<td>1 g <em>per os</em></td>
</tr>
<tr>
<td></td>
<td>Clindamycin</td>
<td>600 mg <em>per os</em></td>
</tr>
</tbody>
</table>

According to the French Infectious Diseases Society (SPILF), 2002.

► **Prevention of rheumatic fever (RF)**

Antibiotic prophylaxis for rheumatic fever is indicated in patients up to the age of 25, who have had a rheumatic fever, with or without cardiac involvement.

► **Cardiovascular drugs**

No treatment is effective in delaying the indication of surgery.

Beta blockers are indicated for Marfan syndrome to prevent aortic dissection. They may be used for ascending aorta dilation (off-label).

In the case of HF, causal treatment is the primary approach (surgery). Medical treatment must follow the usual guidelines (cf. *Systolic heart failure* guide).
In the case of right HF, treatment with diuretics must combine a loop diuretic (furosemide) and a potassium-sparing diuretic (spirolactone).

► **Antiarrhythmic treatment of AF**
See *Atrial fibrillation* ALD guide.

► **Cardioversion (AF)**
Cardiac rhythm disorders occur particularly frequently and are poorly tolerated in patients with severe valvular heart disease and complex congenital heart disease.
Sinus rhythm (SR) should be restored in the case of valvular or congenital heart disease when surgery is not considered.
There is no indication for attempting to reduce atrial fibrillation (AF) in patients with MS prior to surgery. Invasive treatment must be discussed when AF occurs in patients with a mitral valve condition or congenital heart disease.
Cardioversion is considered in the case of persistent post-operative AF during the third month assessment.

► **Treatment of polyglobulia**
This is required for certain complex cyanogenic congenital heart diseases.
In the case of major (haematocrit > 65%) or symptomatic PG, bloodletting is used. Sometimes bloodletting may be combined, based on expert opinion, with a treatment using hydroxyurea or pipobroman to slow down the production of red blood cells by the marrow (off-label).

► **Treatment of PAH**
Cyanogenic congenital heart diseases with highly symptomatic pulmonary artery hypertension (PAH) may benefit from the medical treatments for PAH (bosentan, sildenafil).

► **Contraception and pregnancy**
Women with a valvular disease must discuss the type of contraception they should take with a gynaecologist and cardiologist, taking into account their proper risk of thromboembolism and their risk of infection (IUD is contraindicated for women with a high or intermediate risk of infectious endocarditis).
Any pregnancy with a prosthetic mechanical valve entails a high risk, whatever anticoagulant treatment procedures are being used. The anticoagulant treatment management must involve a cardiologist, an obstetrician and anaesthetist. It is based on VKAs or heparin. VKAs entail a risk of embryopathy in around 5% of cases between six and nine weeks of gestation and a risk of cerebral foetopathy in 1 to 2% of cases beyond this period. But they offer the mother greater safety. Heparin entails an increased
risk of thrombosis and a high mortality rate for the mother, which could be linked to an unsuitable dosage of heparin or inadequate monitoring of the anticoagulation level, especially when switching between VKAs and heparin. During the first trimester, the choice between heparin and VKAs must take into account the patient’s wishes, compliance to treatment and the possibility of using low doses of VKAs. Continuing with a VKA (with the same INR) can be considered when treatment with heparin could entail a higher individual risk of thromboembolism than with VKAs. Switching to heparin requires increased monitoring of the anticoagulation level and platelet count.

VKAs are recommended during the second and third trimesters, until the 36th week, the period during the pregnancy when heparin is required, and then VKAs are resumed after the delivery. The target INR remains unchanged.

Treatment with heparin must be monitored closely. For non-fractionated heparin (NFH): APTT between 2 and 3 times the control level or anti-Xa activity between 0.35 and 0.7; for LMWH: anti-Xa activity between 0.7 and 1.2 U/ml.

The prenatal diagnosis will be adapted to the period of intra-uterine exposure to VKAs.

► **Influenza and pneumococcal vaccination**

Vaccination is recommended.

### 2.5 Invasive treatment

The indication for surgical or percutaneous intervention must be considered in symptomatic patients with severe valvular heart disease.

The indication should also be considered in asymptomatic patients after a risk assessment in order to establish the benefit/risk ratio (spontaneous progression versus early intervention).

The risk assessment is a key element to determine the type of treatment; it is made by the specialized medical and surgical team. The risk of surgery is linked to age, comorbidities (respiratory failure, renal failure, coronary lesions) and to the complexity of the procedure being considered (combined procedures). Euroscore is used to evaluate the risk of surgery (www.euroscore.org).

In the case of congenital heart disease, the risk of surgery is related to the number of operations the patient has undergone previously, as well as to the level of PG and PAH. Specialized teams are required to perform surgery in the case of complex and highly complex congenital heart diseases.

If surgery is the option, it should be the least invasive possible: percutaneous mitral commissurotomy (PMC) in the case of MS, conservative valve surgery in the case of MR. Valve replacement still remains the norm for the aortic orifice.
In the case of congenital heart disease, pulmonary valve replacement should be considered depending on the impact of the pulmonary insufficiency on the RV.

If surgery is not an immediate option, the medical and surgical team must decide beforehand what change will be the threshold leading to surgery.

► Choosing the type of prosthetic valve

Bioprostheses do not require long-term anticoagulant treatment, but have a more restricted lifetime than mechanical prostheses. The choice of prosthesis is determined by age, life expectancy, comorbidities, contraindications to treatment with VKAs and the patient’s informed choice.

► Valve conservative strategy

In the case of severe organic MR, mitral valve repair is the preferred option if feasible.

In the case of MS, percutaneous mitralcommissurotomy (PMC) remains the preferred intervention when the anatomy is favourable.

In the case of AS and AR, surgical aortic valvuloplasty has an important role to play in the treatment of adolescents and young adults, but is very limited afterward.

In functional MR due to LV dysfunction, the indication of surgery is more restrictive than with organic MR.

► Percutaneous treatment of valvular heart disease

PMC is an effective method if performed by a well-trained team. It is indicated for symptomatic patients and may be considered in asymptomatic patients because of the low interventional risk in order to reduce the risk of thromboembolism.

In the case of AS, indications for percutaneous aortic valvuloplasty are rare, except in the treatment of adolescents and young adults.

Balloon dilation of pulmonary valve stenosis is a first-line procedure in adults, using suitable techniques.

Percutaneous implantation of prosthetic valves in the aorta or pulmonary area is currently being assessed.

► Repeat surgery

Repeated surgery is a major risk. It is particularly frequent for congenital heart diseases.

The deterioration of a bioprosthesis gives rise to the discussion of repeated surgery.
The desinsertion of a prosthesis is managed on a case-by-case basis. In the absence of infectious endocarditis, repeated surgery is only option when the leak is large or complicated by significant haemolysis. Thrombosis of a valve prosthesis is a rare, but serious complication, affecting mechanical prostheses in particular. Obstructive thromboses are a life-threatening surgical emergency. Infectious endocarditis of a prosthesis is a serious complication often requiring surgery.

► Associated coronary revascularisation

In the case of significant coronary lesions, coronary revascularisation is generally recommended. Coronary patients are older and more symptomatic, and often have impaired LV function. Associated coronary disease increases the risk of surgery, even if there is no associated surgical coronary procedure.

2.6 Procedure in the event of non-cardiac surgery

► Risks associated with heart disease

Assessing the risk of non-cardiac surgery is particularly important in the case of severe valvular heart disease, as it may be a major risk factor in perioperative cardiovascular complications. In the case of patients with valvular heart disease, the risk assessment must take into account the patient’s age, symptoms, the occurrence of rhythm disorders, severity and type of valvular heart disease, left ventricular function, pulmonary artery pressure and comorbidities, especially coronary disease.

Aortic stenosis (AS) is the most exposed to perioperative complications, irrespective of the patient’s characteristics.

Patients with a congenital heart disease and PAH are most at risk of life-threatening complications after non-cardiac surgery and the indication must carefully discussed. It requires an appropriate anaesthesia.

► Type of non-cardiac surgery.

The risk depends on the type of surgery and, among the general population, three levels of risk are usually identified.
RISK OF CARDIOVASCULAR COMPLICATIONS* WITH NON-CARDIAC SURGERY

High-risk procedures (cardiac risk)
– major urgent procedures, particular with elderly patients
– aortic surgery and other major vascular surgery
– peripheral vascular surgery
– surgery required long term, associated with volume variations and/or major blood loss

Intermediate-risk procedures
– carotid endarterectomy
– head and neck surgery
– intraperitoneal and thoracic surgery
– orthopaedic surgery
– prostate surgery

Low-risk procedures
– endoscopic procedures
– superficial surgery
– cataract surgery
– breast surgery

*Deaths caused by cardiac complications and non-fatal myocardial infarction.


► Anticoagulant treatment

Procedures with a low risk of bleeding (dental treatment, including extractions), must be performed with no modification of the oral anticoagulant treatment.

In the case of surgical or diagnostic procedures with a high risk of bleeding, anticoagulant treatment with VKAs must be stopped 3-4 days before.

The decision not to switch to heparin may be considered if the risk of embolism is low, and only with the cardiologist’s consent.

In high-risk patients, especially those with a mechanical prosthetic valve, switching to heparin is essential.

2.7 Implantable medical devices

These treatments require a specialized cardiological management, particularly in the case of congenital heart disease.

► Pacemakers

If indicated.
► Implantable defibrillators
If indicated.

► Ventricular resynchronisation
Biventricular resynchronisation, if indicated, must be carried out by experienced medical teams.
3. Follow-up

Follow-up is based on a clinical and echocardiographic examination and blood tests where anticoagulant treatment is involved.

3.1 Objectives

For all patients:
- Provide the best possible treatment and improve quality of life
- Prevent infectious endocarditis
- Prevent thromboembolism
- Prevent and screen for bleeding complications if anticoagulant treatment
- Improve education of the patients and their families on lifestyle and dietary measures, treatment procedures, their risks and monitoring.

For non-operated patients:
- Detect new symptoms
- Monitor the progression of valvular lesions and their myocardial impact
- Perform surgery at the right time

For operated patients:
- Make sure that there is no thromboembolic complication if the patient has a prosthetic valve
- Detect signs of degeneration of bioprostheses
- Make sure that there is no desinsertion of prostheses.
- Detect early deterioration in PMC or valvuloplasty

3.2 Professionals involved

The follow-up of patients with severe valvular heart disease is under the responsibility of the primary care physician and/or cardiologist and the specialized team, in the case of complex congenital heart disease. A visit to the general practitioner is recommended at least every year for stable, asymptomatic patients, with no medication. The biochemist is involved in the case of anticoagulant treatment, as well as the coagulant specialist when achieving a balanced anticoagulant treatment is extremely difficult.
► Referal to specialized management

- Cardiologist
  A visit with a clinical and echocardiographic assessment is recommended at a frequency varying according to the degree of severity and the potential progression of the valvular heart disease (every 6 months to 3 years).
  In the case of severe non operated valvular heart disease, a visit is recommended every 6 to 12 months. If the condition is less severe, a consultation every 1 to 3 years is sufficient.
  When a valvular heart disease had undergone successful surgery, a baseline post-operative visit should be performed (involving a clinical assessment, echocardiography and blood tests) 2 or 3 months after surgery, followed by an annual clinical visit.
  Consultations with the specialized team must be held regularly for patients with congenital cardiac disease, especially complex conditions.

- Other professionals may be involved, depending on the medical and social setting:
  - surgeon and interventional cardiologist
    - if repeated surgery is being considered
  - cardiovascular rehabilitation cardiologist
    - if the patient’s clinical condition requires rehabilitation
  - dietician, in the case of obesity
    (*Legislation does not provide for reimbursement of the services of a dietician.*)
  - physiotherapist, after surgery

► Multidisciplinary management

Management involving a educational programme and/or specific coordinated follow-up is recommended, especially if anticoagulant treatment has been prescribed and for patients with complex or highly complex congenital heart diseases, or with complications (HF, rhythm disorders which have already required hospitalisation).

► In and out-hospital collaboration

Management of the return home:
- patient is discharged with a discharge plan, only once his/her condition is stable
- primary care physician is informed of the management plan and arranges follow-up treatment;
- Patients and their families are given precise instructions on the aspects to be monitored after they return home
Preventing emergency hospitalisation by identifying and remedying:
- Medical complications (HT, infections, anaemia, renal failure etc.)
- Lack of social support etc.
- Poor compliance with treatment, lifestyle or dietary measures etc;
- Poorly planned discharge from hospital
Prevention of emergency hospitalisations through surveillance and assistance for the following:

Prevention of emergency hospitalisations through surveillance and assistance for the following:
- Acute HF
- Rhythm disorders (congenital heart disease)
- Suspected infectious endocarditis
- Suspected thrombosis of prosthetic valve or embolic complications
- Haemorrhagic complications or overdosing of VKAs

3.3 Clinical monitoring

► Interview
This clarifies the activities of daily living, while looking for and quantifying symptoms, particularly dyspnoea.
It looks for thromboembolic or haemorrhagic complications.
It helps to verify how much the patient knows about prophylactic measures for infectious endocarditis and the rules for monitoring anticoagulant treatment, if relevant.

► Clinical examination
The schedule and frequency of visits must be established on the basis of the initial assessment.
Any suspected prosthetic thrombosis requires immediate management by a specialist medical and surgical team.
Endocarditis must be considered in the case of unexplained fever and an emergency visit is necessary. Any suspicion of infectious endocarditis requires immediate management by a specialized medical and surgical team.
A cerebral abscess must be considered in patients with a cyanogenic heart diseases, if they have an isolated fever or neurological symptoms.

3.4 Additional tests

► TTE
This must be performed by an experienced echocardiography technician:
● Severe non operated valvular heart diseases (every 6 to 12 months)
● Less severe valvular heart diseases (every 2 to 3 years)
● Prosthetic valves and plasties:
  ‣ if bioprosthesis, baseline post-operative TTE, then every year from
    the fifth year
  ‣ if any clinical change
  ‣ at least every three years for patients in a stable condition with
    mechanical prostheses or plasties
● Complex congenital heart disease

A regular quantitative assessment of left ventricular function is mandatory in
the case of AI and MI, where LV dysfunctions are not always preceded by
symptoms.

▶ Additional examinations
● ECG, as part of the visit to the cardiologist
● Holter, regularly with certain congenital heart diseases
  ‣ if an inaugural sign is noted during the interview or clinical examination
● Chest X-ray
  ‣ If any new symptom occurs

▶ Laboratory tests
● INR if anticoagulant therapy:
  ‣ 1-2 times a week until the value has stabilised
  ‣ Once a month at least after

● If a new symptom and depending on the treatment:
  ‣ Full blood count with investigation for schizocytes
  ‣ Blood sodium, potassium
  ‣ Serum creatinine and calculation of creatinine clearance (Cockroft
    and Gault formula)
  ‣ Liver function tests
  ‣ LDH
Appendix 1. Participants

This study was coordinated by Dr Nikita de Vernejoul, project leader at the department of Long-Term Conditions and Conventional Agreements, and carried out by the following participants:

- Dr Luc Darnige, biochemist, specialized in haematology and coagulation, Paris
- Professor Guy Durand de Gevigney, cardiologist, Lyon
- Dr Sylvie Gillier, general practitioner, Saint-Sébastien-sur-Loire
- Dr Laurence Iserin, cardiologist, Paris
- Professor Bernard Iung, cardiologist, Paris
- Professor Alain Leguerrier, cardiac surgeon, Rennes
- Professor Jean-François Obadia, cardiac surgeon, Lyon
- Dr Remi Pécaud, CNAMTS
- Dr Sébastien Ducourant, RSI consultant

The guide has also been reviewed by representatives of AFDOC (French National Federation of Cardiovascular Surgery and Disease Patients) and Afssaps.
Appendix 2. Target INR in the case of a mechanical prosthesis

The target INR in the case of a mechanical prosthetic valve depends on the type of prosthesis and the risks related to the patient.

<table>
<thead>
<tr>
<th>Thrombogenicity of the prosthesis</th>
<th>Risks related to the patient</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>No risk factor</td>
</tr>
<tr>
<td>Low</td>
<td>Target 2.5</td>
</tr>
<tr>
<td></td>
<td>Range 2 to 3</td>
</tr>
<tr>
<td>Moderate</td>
<td>Target 3.0</td>
</tr>
<tr>
<td></td>
<td>Range 2.5 to 3.5</td>
</tr>
<tr>
<td>High</td>
<td>Target 3.5</td>
</tr>
<tr>
<td></td>
<td>Range 3 to 4</td>
</tr>
</tbody>
</table>

Adapted from the European Society of Cardiology, 2007.

The risks related to the patient are:
- History of a thromboembolism, AF
- LA > 50 mm, intra-atrial dense contrast
- EF < 35%
- State of hypercoagulability

The factors related to valvular involvement are:
- Mitral, tricuspid, pulmonary location
- MS
Appendix 3. Self-measurement and self-monitoring in the case of VKA treatment

**Self-measurement** (measuring INR from a capillary blood sample) is considered if:
- The patient is physically capable of carrying out this procedure and has the mental and cognitive capacity to acquire knowledge or, if this is not the case, a close relative may take the measurement.
- An adequate training programme is set up to educate patients and/or the persons caring for them.
- The self-testing device is regularly checked using a quality control programme.

**Self-monitoring** of anticoagulation therapy (dose adjustment according to INR by patients themselves) may be subsequently considered if the patients correctly perform self measurement and if they have the necessary mental and cognitive capacities to learn about and perform this dose adjustment or if one of their relatives can perform it.

In every case, the ability of patients and/or their relatives to conduct this self-measurement and self-monitoring must be checked regularly.
### Appendix 4. Management of valvular heart diseases

#### Aortic stenosis (AS)

<table>
<thead>
<tr>
<th><strong>Introduction</strong></th>
<th>The most frequent valvular heart disease</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>— CAS: Calcified AS in elderly patients</td>
</tr>
<tr>
<td></td>
<td>— Congenital AS in the youngest patients</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th><strong>Clinical assessment</strong></th>
<th>Systolic murmur and absence of S2</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Dyspnoea, angina, fainting</td>
</tr>
<tr>
<td></td>
<td>Severe AS: every 6-12 months</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th><strong>TTE</strong></th>
<th>Investigation of choice:</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>— diagnosis and degree of stenosis</td>
</tr>
<tr>
<td></td>
<td>— parietal thickness, size and function of the LV</td>
</tr>
<tr>
<td></td>
<td>— new assessment if signs or symptoms have changed</td>
</tr>
<tr>
<td></td>
<td>— haemodynamic changes and LV function in pregnant women with AS</td>
</tr>
</tbody>
</table>

**Follow-up:**
- Every 6-12 months if severe unoperated AS
- Every 1-2 years if less severe AS

<table>
<thead>
<tr>
<th><strong>Exercise test</strong></th>
<th>— May be useful for asymptomatic patients to highlight symptoms and an abnormal BP profile on exercise</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>— Contraindicated for symptomatic patients</td>
</tr>
</tbody>
</table>

| **Coronary angiography** | — Recommended before an aortic valve replacement |

| **Surgical indications for valve replacement** | — Severe symptomatic AS (dyspnoea NYHA classes II-III-IV, angina) |
|                                                | — Severe AS with another indication for cardiac surgery (coronary artery bypass surgery, ascending aorta surgery or another cardiac valve) |
|                                                | — Severe asymptomatic AS and LV dysfunction |
|                                                | — Severe asymptomatic AS and a pathological exercise test |
|                                                | — Severe asymptomatic AS with moderate to severe calcification and a speed of progression of the velocity peak $\geq 0.3$ m/s per year |
|                                                | — Low-gradient AS ($< 40$ mmHg) and LV dysfunction |
# Aortic stenosis (AS)

<table>
<thead>
<tr>
<th>Surgical treatment</th>
<th>Valve replacement</th>
<th>Mechanical valve</th>
</tr>
</thead>
<tbody>
<tr>
<td>In favour of a mechanical prosthesis:</td>
<td>Patient’s informed choice, provided there is no long-term contraindication for anticoagulant treatment</td>
<td>— Age &lt; 65-70 and long life expectancy</td>
</tr>
<tr>
<td></td>
<td>— Risk of accelerated valvular deterioration</td>
<td>— Patients already taking VKAs for another mechanical prosthesis</td>
</tr>
<tr>
<td></td>
<td>— Patients for whom repeat surgery would entail considerable risk</td>
<td>— Patients for whom repeat surgery would entail considerable risk</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Surgical treatment</th>
<th>Valve replacement</th>
<th>Bioprosthesis</th>
</tr>
</thead>
<tbody>
<tr>
<td>In favour of a bioprosthesis:</td>
<td>Patient’s informed choice</td>
<td>Elderly patients &gt; 65-70 years</td>
</tr>
<tr>
<td></td>
<td>— Elderly patients &gt; 65-70 years</td>
<td>— Absolute contraindication or contraindication linked to VKAs</td>
</tr>
<tr>
<td></td>
<td>— Repeated surgery for thrombosis of a mechanical valve in patients with poorly balanced anticoagulant treatment</td>
<td>— Patients for whom repeat surgery would entail a slight risk</td>
</tr>
<tr>
<td></td>
<td>— Patients with a reduced life expectancy or severe comorbidities</td>
<td>— Women wishing to become pregnant</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Indication for interventional cardiology</th>
<th>Aortic balloon valvuloplasty considered for:</th>
<th>Young adult patients with congenital AS</th>
</tr>
</thead>
<tbody>
<tr>
<td>Medication</td>
<td>Prevention of infectious endocarditis</td>
<td></td>
</tr>
</tbody>
</table>

Severe AS: aortic area < 0.6 cm²/m² or < 1.0 cm².
# Aortic Regurgitation (AR)

## Introduction
Most frequent causes: dystrophy, bicuspid aortic valve

## Clinical assessment
**Diastolic murmur, pulse rate, BP**

**Follow-up:**
- Severe unoperated AR: every 6-12 months

## TTE
**Investigation of choice:**
- size of the leak associated with the AR and cause of the AR (valve morphology, size and morphology of aortic root)
- parietal thickness, size and function of the LV

**Follow-up:**
- Severe AR: every 6-12 months
- Moderate AR: every 2-3 years (once a year if initial aortic dilation)

## Isotopic angiography
- If TTE inconclusive or in the case of LV systolic dysfunction

## Exercise test
- If symptoms inconclusive

## MRI
- Assessing the aortic root, AR and LV function, if TTE inconclusive

## Coronary angiography
- Before an aortic valve replacement in patients at risk of coronary heart disease

## Surgical indications for valve replacement
**Severe AR**
- Symptomatic patients (dyspnoea NYHA classes II-III-IV, angina)
- Asymptomatic patients with EF <50% at rest or EF at rest > 50% and severe dilation of LV
- Patients with another indication for cardiac surgery (coronary artery bypass, ascending aorta surgery or another valve replacement)

**AR of any severity**
- Patients with ascending aortic dilation (maximum diameter > 55 mm, > 50 mm if bicuspid aortic valve, > 45 mm if Marfan syndrome)

## Surgery
- Valvular procedure
  - Mechanical valve
  - Bioprosthesis
  - Valve repair
- Surgery on ascending aorta

## Interventional cardiology
- No
## Aortic Regurgitation (AR)

| Medication | — No effective medication as an alternative to surgery  
| — Prevention of infectious endocarditis |
| Specific subgroup | — Annual assessment of the size of the ascending aorta (if diameter > 45 mm)  
| Bicuspid aortic valve with ascending aortic dilation | — Ascending aortic surgery if diameter > 50 mm or if the diameter increases by > 5 mm/year  
| | — If aortic valve replacement for severe AR or AS, ascending aortic surgery is indicated if the diameter of the ascending aorta is > 45 mm  
| | — Treatment with beta blockers recommended in the case of a bicuspid aortic valve and ascending aortic dilation (diameter > 45 mm) without any surgical indication |
# Mitral Regurgitation (MR)

## Introduction
Organic MR is degenerative in most cases, more rarely rheumatic. Ischaemic or functional MI differs in its prognosis and surgical indications.

## Clinical assessment
Follow-up:
- Severe unoperated MR: every 6-12 months

## TTE
Investigation of choice:
- Size and function of the LV, RV and the size of the LA, PAP
- Mechanism of the leak and the severity of MR
- Tricuspid involvement
Follow-up:
- Severe MI: 6-12 months
- Less severe MI: every 2-3 years

## TOE
- Before surgery, to assess the extent of MI when the TTE has not provided sufficient information about the severity of MR, its mechanism and the possibilities of plasty
- After surgery if plasty

## Exercise test
To assess how exercise is tolerated

## Coronary angiography
- Before mitral surgery in patients at risk of coronary heart disease

## Surgical indications
- Symptomatic severe organic MR
- Asymptomatic severe organic MR with LV dysfunction (EF < 60%), AF or PAH
- Severe organic MR with EF > 60% and high probability of plasty

## Surgical treatment

### Valvuloplasty
Preferred treatment
Post-operative baseline assessment after 3 months: ECG, echocardiogram, laboratory tests
Anticoagulant treatment/aspirin for 3 months after plasty

### Valve replacement
- Mechanical valve
- Bioprosthesis
Post surgery: ECG, chest X-ray, echocardiogram
Treatment with anticoagulants

## Interventional cardiology
- No
## Mitral Regurgitation (MR)

<table>
<thead>
<tr>
<th>Medication</th>
<th>No effective treatment with medication as an alternative to surgery</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>VKAs in the case of AF</td>
</tr>
<tr>
<td></td>
<td>Cardioversion pointless in the case of severe MI due to be operated on</td>
</tr>
<tr>
<td></td>
<td>Prevention of infectious endocarditis</td>
</tr>
</tbody>
</table>

### Specific subgroups

**Ischaemic MR**

Indications for surgery:

- Patients with severe MR, EF < 30%, due to have bypass surgery
- Patients with less severe MR, due to have bypass surgery if plasty is feasible
- Patients with severe symptomatic MR with EF ≥ 30% and option of revascularisation
- Patients with severe MR with EF ≥ 30% without option of revascularisation, resistant to treatment with medication and with low comorbidity

**Functional MR**

Medical treatment of heart failure
Ventricular resynchronisation considered
Annuloplasty sometimes indicated
# Mitral stenosis (MS)

<table>
<thead>
<tr>
<th><strong>Introduction</strong></th>
<th>Predominant rheumatic aetiology</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Clinical assessment</strong></td>
<td>Follow-up:</td>
</tr>
<tr>
<td></td>
<td>— Severe unoperated MS: every 6-12 months</td>
</tr>
<tr>
<td><strong>TTE</strong></td>
<td>Investigation of choice:</td>
</tr>
<tr>
<td></td>
<td>— diagnosis and degree of stenosis (average gradient, mitral area, PAP)</td>
</tr>
<tr>
<td></td>
<td>— other valvular lesions and determining valve morphology (to assess the possibility of percutaneous commissurotomy)</td>
</tr>
<tr>
<td></td>
<td>Follow-up:</td>
</tr>
<tr>
<td></td>
<td>— Severe non operated MS: every 6-12 months</td>
</tr>
<tr>
<td></td>
<td>— Less severe MS: every 2-3 years</td>
</tr>
<tr>
<td><strong>TOE</strong></td>
<td></td>
</tr>
<tr>
<td></td>
<td>— To investigate left atrial thrombi and define more clearly the anatomy of the MS in patients being considered for a percutaneous commissurotomy</td>
</tr>
<tr>
<td></td>
<td>— TTE inconclusive</td>
</tr>
<tr>
<td><strong>Exercise test</strong></td>
<td>— If symptoms inconclusive</td>
</tr>
<tr>
<td><strong>Interventional indications</strong></td>
<td>Symptomatic severe MS:</td>
</tr>
<tr>
<td></td>
<td>— PMC as first-line treatment</td>
</tr>
<tr>
<td></td>
<td>— Surgery if PMC contraindicated</td>
</tr>
<tr>
<td></td>
<td>Asymptomatic severe MS:</td>
</tr>
<tr>
<td></td>
<td>— PMC if high thromboembolic risk</td>
</tr>
<tr>
<td></td>
<td>— If wish to become pregnant</td>
</tr>
<tr>
<td><strong>Valve surgery</strong></td>
<td>— Mechanical valve</td>
</tr>
<tr>
<td></td>
<td>— Bioprosthesis</td>
</tr>
<tr>
<td></td>
<td>— Open-heart commissurotomy</td>
</tr>
<tr>
<td><strong>Medication</strong></td>
<td>— No effective treatment with medication as an alternative to surgery</td>
</tr>
<tr>
<td></td>
<td>— SR maintenance</td>
</tr>
<tr>
<td></td>
<td>— Anticoagulant treatment if:</td>
</tr>
<tr>
<td></td>
<td>- MS in AF (paroxystic, persistent or permanent)</td>
</tr>
<tr>
<td></td>
<td>- history of embolism, even if SR present</td>
</tr>
<tr>
<td></td>
<td>- left atrial thrombus</td>
</tr>
<tr>
<td></td>
<td>- considered when dilation of LA and/or spontaneous contrasts with TTE/TOE</td>
</tr>
</tbody>
</table>

Severe mitral stenosis (MS): MITRAL AREA < 0.9 cm²/m² or < 1.5 cm².
## PROSTHETIC VALVES

| **Introduction** | Population is large and at risk  
| | 75% of complications encountered are associated with prostheses:  
| | — Degeneration in bioprostheses  
| | — Thromboembolism  
| | — Bleeding linked to anticoagulant treatment  
| | — Infectious endocarditis  

| **Clinical assessment** |  
| | — Initial baseline cardiological assessment,  
| | 3 months after surgery, with chest X-ray, TTE, full blood count and INR  
| | — Annual cardiological follow-up to look for prosthetic or ventricular dysfunction, with a complication or progression of the disease affecting another valve  

| **TTE** | Investigation of choice:  
| | — In the event of any clinical change  
| | — At least every 3 years for patients who remain stable  
| | — In the case of a bioprosthesis, TTE every year from the fifth year  

| **Medication** | — Treatment with anticoagulants patients with Mechanical prostheses must take anticoagulant therapy for the rest of their lives. Bioprostheses or a plasty with another indication for anticoagulant therapy (AF, HF with LV dysfunction FE< 30%)  
| | Three months after surgery for bioprosthesis or plasty  
| | Antiplatelet agents must be added to the anticoagulant therapy only for selected patients  
| | — Antibiotic prophylaxis to prevent infectious endocarditis  

## Appendix 5. Management of complex congenital heart diseases

### Example of operated tetralogy of Fallot

<table>
<thead>
<tr>
<th><strong>Introduction</strong></th>
</tr>
</thead>
</table>
| The most frequent of the complex congenital heart diseases  
The survival rate after surgery is excellent |

<table>
<thead>
<tr>
<th><strong>Clinical assessment</strong></th>
</tr>
</thead>
</table>
| — Follow-up once a year if residual lesions: PR/PS, TR, AR, VSD  
otherwise, every 2 years |
| — ECG once a year or every 2 years, looking for frequent rhythm disorders, especially atrial |

<table>
<thead>
<tr>
<th><strong>TTE</strong></th>
</tr>
</thead>
</table>
| Once a year if residual lesions or every 2 years  
TOE if necessary |

<table>
<thead>
<tr>
<th><strong>Holter</strong></th>
</tr>
</thead>
<tbody>
<tr>
<td>— May be useful for symptomatic patients or those showing signs of haemodynamic distress to highlight rhythm disorders</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th><strong>Cardiac catheterisation</strong></th>
</tr>
</thead>
</table>
| — Before surgery, if residual lesions, in order to determine the anatomy of the coronary arteries before implanting a stent in the pulmonary artery  
— If endovascular implantation of a pulmonary valve |

<table>
<thead>
<tr>
<th><strong>Exercise test</strong></th>
</tr>
</thead>
<tbody>
<tr>
<td>— May be useful for determining performance during exercise and screening for arrhythmias</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th><strong>Chest X-ray</strong></th>
</tr>
</thead>
<tbody>
<tr>
<td>— Sometimes useful if cardiomegaly</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th><strong>MRI</strong></th>
</tr>
</thead>
<tbody>
<tr>
<td>— To evaluate RV function and size, as well as PR</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th><strong>Surgical indications</strong></th>
</tr>
</thead>
</table>
| — Stenosis on right pathway  
— AR  
— Residual VSD, severe PR (with symptoms and/or major dilation of RV) |

<table>
<thead>
<tr>
<th><strong>Surgical treatment</strong></th>
</tr>
</thead>
</table>
| — Surgery on residual lesions  
— Rhythmic surgery |

<table>
<thead>
<tr>
<th><strong>Interventional cardiology</strong></th>
</tr>
</thead>
</table>
| — Angioplasty and stent for pulmonary arteries  
— Endocavitary implantation of a pulmonary valve  
— Pulmonary balloon valvuloplasty with percutaneous closure of residual VSD |
Example of operated tetralogy of Fallot

<table>
<thead>
<tr>
<th>Medication</th>
</tr>
</thead>
<tbody>
<tr>
<td>— No effective treatment with medication as an alternative to surgery</td>
</tr>
<tr>
<td>— SR maintenance</td>
</tr>
<tr>
<td>— Antiarrhythmic treatment (beta blockers, amiodarone)</td>
</tr>
<tr>
<td>— Prevention of infectious endocarditis</td>
</tr>
</tbody>
</table>
Appendix 6 Indications for coronary angiography

<table>
<thead>
<tr>
<th>Indications for coronary angiography in patients with valvular heart disease</th>
</tr>
</thead>
<tbody>
<tr>
<td>Prior to valve surgery in patients with severe valvular heart disease and:</td>
</tr>
<tr>
<td>— a history of coronary disease</td>
</tr>
<tr>
<td>— suspected ischaemic heart disease</td>
</tr>
<tr>
<td>— LV dysfunction</td>
</tr>
<tr>
<td>— men aged &gt; 40 years and menopausal women</td>
</tr>
<tr>
<td>≥ 1 CV-RF</td>
</tr>
<tr>
<td>When coronary disease is likely to be responsible for severe MI (ischaemic MI)</td>
</tr>
</tbody>
</table>
Appendix 7. References


