



## **Guidelines: Congenital Aortic Valve Stenosis**

**PD Dr. med. Peter Ewert**

Senior Physician, Department of Congenital Heart Disease / Pediatric Cardiology,  
Deutsches Herzzentrum Berlin

# **Guidelines: Congenital Aortic Valve Stenosis**

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**Authors:** Peter Ewert, Berlin; Nikolaus Haas, Bad Oeynhausen

- 1) Disease: Aortic valve stenosis**
- 2) Definition – Classification – Basic information**
- 3) Identifying symptoms**
- 4) Diagnostic imaging**
- 5) Treatment**
- 6) Follow-up**
- 7) Prevention**

# 1) Disease

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## **Aortic valve stenosis**

Defined as: isolated aortic valve stenosis in children and adolescents

## 2) Definition – Classification – Basic information

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Aortic valve stenosis is used to mean narrowing of the aortic valve. This is caused by incomplete opening of the semilunar valve or by the valve ring being too small. A combination of these is possible. The exact morphology of the valve is important for the therapy. Frequently the valve is tricuspid but the cusps are fused at the commissures. Individual cusps may be only rudimentary or completely missing, so that bicuspid valves result. Further, there may be dysplasia with thickened, less mobile cusps. In severe forms the valve structure may be almost completely lacking and instead resemble a perforated (monocuspid) membrane.

# Division of aortic valve stenoses according to severity

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	Heart catheter peak-to-peak gradient	cw Doppler V <sub>max</sub> <i>ACC/AHA</i> <i>ESC</i>	Bernoulli peak instantaneous gradient	Bernoulli mean instantaneous gradient <i>ACC/AHA</i> <i>ESC</i>	Echo aortic valve area <i>ACC/AHA</i> <i>ESC</i>
Trivial					
Slight	< 30 mmHg	< 3 m/s	< 36 mmHg	< 25 mmHg	> 1.5 cm <sup>2</sup> (> 1 cm <sup>2</sup> /m <sup>2</sup> )
Moderate	30-50 mmHg	3-4 m/s	36-64 mmHg	25-50 mmHg	1-1.5 cm <sup>2</sup> (0.6-1 cm <sup>2</sup> /m <sup>2</sup> )
Severe	> 50 mmHg	> 4 m/s	> 64 mmHg	> 50 mmHg	< 1 cm <sup>2</sup> (< 0.6 cm <sup>2</sup> /m <sup>2</sup> )

# **“Critical” aortic valve stenosis**

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**What is a critical aortic valve stenosis?**

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Functionally, critical aortic valve stenosis is defined as severe stenosis in neonates that would lead to decompensation of the left ventricle if not treated.

Systemic perfusion is dependent on the ductus, i.e. the stenosis is so severe that additional perfusion of the circulation through an open ductus is necessary to sustain life.

# “Critical” aortic valve stenosis

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**=> Task of authors: clarify definition of “critical” with reference to the literature**



# “Critical” aortic valve stenosis

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=> Task of authors: clarify definition of “critical” with reference to the literature

The “old” guidelines said:

Definition:            *“in critical aortic stenosis in neonates...”*

Main symptoms:    *“in critical aortic stenosis in neonates and young infants...”*

# **“Critical” aortic valve stenosis**

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**=> Task of authors: clarify definition of “critical” with reference to the literature**

***We have the following definitions:***

***... critical aortic stenosis as defined by ductus dependence***

***... critical stenosis in newborns (in the first 4 weeks of life)***

***... critical aortic stenosis in infants, manifesting in the first 6 weeks of life (Bühlmeyer)***

***... critical stenosis in infants***

## 2) Definition – Classification – Basic information

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### **“Critical” aortic valve stenosis**

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## **2) Definition – Classification – Basic information**

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### **“Critical” aortic valve stenosis**

These stenoses already exist intrauterinely and may therefore be accompanied by endocardial fibroelastosis (a special form of restrictive cardiomyopathy). Very severe stenoses may lead to prenatal hypoplasia of the left ventricle, developing into hypoplastic left heart syndrome.

### 3) Identifying symptoms

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Critical aortic stenosis in the newborn leads to left ventricular insufficiency with drinking weakness, tachydyspnea, tachycardia, pulmonary edema and cardiogenic shock.

If heart function is severely impaired, a heart sound may be lacking.

In patients older than newborns the ejection sound through the left ventricular outflow tract is characteristic. This is transmitted to the carotid vessels and is caused by the turbulent ejection stream from the narrowed valve. This turbulence may be palpable as fremitus in the jugulum.

The physical tolerance of strain may be very good despite severe stenosis and is therefore unsuitable as an identifying symptom.

Severe stenoses can, however, become symptomatic and lead to sudden cardiac death. A severe stenosis is regarded as symptomatic when angina pectoris symptoms occur at rest or during activity, when dyspnea occurs at rest or during mild activity or when syncope occurs.

## **4) Diagnostic imaging**

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**4.1) Aim**

**4.2) Echocardiography**

**4.3) ECG**

**4.4) Long-term ECG**

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**4.5) Ergometry with blood pressure measurement**

**4.6) Heart catheterization**

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**4.7) Cardiac MRT**

**4.8) Cardiac CT**

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**4.9) Performance of diagnostic procedures**

## 4) Diagnostic imaging

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### 4.1) Aim

Confirm diagnosis

Clarify valve morphology

Measure valve ring and LV

Measure pressure gradients

Estimate possibilities of LV compensation

Exclude further disease

Assess prognosis

Plan therapy

## 4) Diagnostic imaging

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### 4.2) Echocardiography

- confirms diagnosis by detailed visualization of the aortic valve apparatus.
- allows differential diagnosis and distinction between AV stenosis and other obstructions below or above the valve.
- provides information on the formation of the valves and their symmetry.
- enables very accurate estimation of additional valve insufficiency.
- also enables quantification of ventricular function by measuring ventricular diameter, ejection fraction (EF) and wall thicknesses.



## 4) Diagnostic imaging

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### 4.2) Echocardiography

In critical aortic valve stenosis LV echocardiography is the most important means for decision-making, e.g. for biventricular or univentricular treatment strategy, primary intervention or operation. Further, echocardiography flanks intensive medical therapy.

Factors playing a role in the evaluation:

aortic valve diameter; LV size and morphology (pointed?); diameter and anatomy of mitral valve; extent of endocardial fibroelastosis.

Before a planned Ross operation the pulmonary valve must be assessed.

## 4) Diagnostic imaging

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### 4.3) ECG

The ECG identifies hypertrophy and repolarization disturbances as signs of myocardial ischemia of the volume-loaded, hypertrophied LV.

### 4.4) Long-term ECG

The long-term ECG identifies ventricular rhythm disturbances and repolarization disturbances during the activities of everyday life.

## 4) Diagnostic imaging

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### 4.5) Ergometry with blood pressure measurement

-is helpful for risk stratification and therapy planning in asymptomatic patients with moderate stenosis.

The following findings are assessed:

- symptoms during exertion
- blood pressure regulation (fall or inadequate rise)
- ventricular rhythm disturbances
- ST changes

**Caution:** In symptomatic patients ergometry is dangerous and therefore counter-indicated.

## **4) Diagnostic imaging**

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### **4.6) Heart catheterization**

- is only indicated if balloon valvuloplasty is planned.

## 4) Diagnostic imaging

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### 4.7) Cardiac MRT

- supplementary to echocardiography and TEE (3D reconstruction), cardiac MRT can be used to visualize the morphology of the aortic valve (e.g. before a planned reconstructive procedure), the pulmonary valve (e.g. before Ross operation) and the coronary arteries.

### 4.8) Cardiac CT

- is not usually necessary. In individual cases it is suitable for coronary visualization but it involves radiation exposure.

## **4) Diagnostic imaging**

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### **4.9) Performance of diagnostic procedures**

- by a pediatric physician qualified in pediatric cardiology
- in a hospital / department for pediatric cardiology.

# Excursus: Guidelines in Germany

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Die AWMF ist das Netzwerk der Wissenschaftlichen Medizinischen Fachgesellschaften in Deutschland.

In der 1962 gegründeten **Arbeitsgemeinschaft der Wissenschaftlichen Medizinischen Fachgesellschaften e.V. (AWMF)** sind derzeit 156 wissenschaftlich arbeitende medizinische Fachgesellschaften organisiert.

Die AWMF berät über grundsätzliche und fachübergreifende Fragestellungen in der wissenschaftlichen Medizin, fördert die Zusammenarbeit ihrer Mitgliedsgesellschaften bei der Wahrnehmung ihrer wissenschaftlich-medizinischen Aufgaben und Ziele sowie den Transfer wissenschaftlicher Erkenntnisse in die ärztliche Praxis. Darüber hinaus vertritt sie in Kooperation mit anderen ärztlichen Organisationen die Interessen der wissenschaftlichen Medizin gegenüber den zuständigen politischen Gremien und der Öffentlichkeit, strebt eine enge Zusammenarbeit mit vergleichbaren Organisationen an und stellt somit einen wichtigen Pfeiler in der medizinischen Organisation Deutschlands dar.

Zentrale Säulen der AWMF sind die Delegierten-Konferenz und das Präsidium. Für die Bearbeitung besonderer Fragestellungen werden Kommissionen aus den Delegierten gebildet und gegebenenfalls Sachverständige aus den Mitgliedsgesellschaften hinzugezogen. Die AWMF organisiert außerdem zwei interdisziplinäre Arbeitskreise.

<http://www.awmf.org/die-awmf.html>

## 5) Therapy

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The only causal therapy is the removal or at least reduction of the stenosis.

If biventricular correction does not seem promising in view of the morphometric data of the left side of the heart (aortic valve diameter, LV size [pointed?], mitral valve diameter and anatomy, extent of endocardial fibroelastosis), univentricular palliation is necessary.

Treatment in children may be surgical or, in suitable cases, by catheter intervention.

Critical aortic valve stenosis also requires supportive therapy.



# **5) Therapy**

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**5.1) Therapy indications**

**5.2) Therapy for critical aortic valve stenosis**

**5.3) Therapy for valvular aortic valve stenosis beyond neonatal age**

**5.3.1) Heart catheter interventions**

**5.3.2) Surgical procedures**

# 5) Therapy

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## 5.1) Therapy indications

Critical aortic stenosis is, by definition, not accompanied by left ventricular failure and requires prompt action after being diagnosed.

In children beyond neonate age the indication for therapy is given if the patient is symptomatic or if the stenosis is severe (indication class I, evidence grade III).

# 5) Therapy

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## 5.1) Therapy indication for severe stenosis

is given

- if Doppler echo flow velocity is over 4m/sec (= calculated instantaneous pressure gradient of over 64 mmHg) or
- if mean flow velocity is over 3.5 m/sec (= calculated mean pressure gradient of over 50 mmHg) or invasively measured peak-to-peak pressure gradient is 50 mmHg.

These values apply to a resting child and to the sufficient LV.

# 5) Therapie

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## 5.1) Therapy indication for moderate stenosis (3-4m/sec)

In moderate stenosis with lower gradients, additional criteria must be taken into account so as not to delay treatment: if LV function is impaired or LV dilatation is present or the resting ECG shows repolarization and the ergometry pathological findings, the indication for therapy is regarded as given, despite lower pressure gradients.

If aortic valve findings are not congruent or LV function is reduced, dobutamine stimulation (e.g. 10mcg/kg as bolus) can help to reveal the true grade of stenosis and distinguish it from a functional impairment of other origin (myocarditis, dilatative cardiomyopathy).

# 5) Therapy

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## 5.2) Therapy for critical aortic valve stenosis

If severe heart failure is present it is advisable to treat it before the valve procedure (catheter intervention, operation with heart-lung machine) to create more stable pre-procedural conditions.

With prostaglandin the systemic perfusion may be supported.

If there is isolated impairment of LV pumping function with dilatated ventricle, inotropic support and loop diuretics are indicated.

In severe cases intensive medical care with artificial ventilation and catecholamines is necessary.

When cardiogenic shock is present, acidosis is additionally treated.

## 5.2) Therapy for critical aortic valve stenosis

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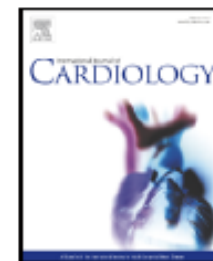
If the valve ring is well developed, catheter balloon dilatation or surgical commissurotomy of the valve are options.

Balloon valvuloplasty is less invasive and shows good long-term results.

Commissurotomy requires a thoracotomy and use of the heart-lung machine. It is therefore more of a strain on the patient, particularly the neonate with unstable circulation.

There are no data documenting the superiority of one procedure over the other. If the valve ring is too small or the valve is very dysplastic and a repair procedure (balloon valvuloplasty, commissurotomy) is not promising, the neonatal Ross operation may be indicated.

If the structures of the left side of the heart are severely hypoplastic, univentricular palliation may be necessary.



## Balloon valvuloplasty in the treatment of congenital aortic valve stenosis – <sup>A</sup> retrospective multicenter survey of more than 1000 patients

P. Ewert <sup>a,\*</sup>, H. Bertram <sup>b</sup>, J. Breuer <sup>c</sup>, I. Dähnert <sup>d</sup>, S. Dittrich <sup>e</sup>, A. Eicken <sup>f</sup>, M. Emmel <sup>g</sup>, G. Fischer <sup>h</sup>, R. Gitter <sup>i</sup>, M. Gorenflo <sup>j</sup>, N. Haas <sup>k</sup>, E. Kitzmüller <sup>l</sup>, A. Koch <sup>m</sup>, O. Kretschmar <sup>n</sup>, A. Lindinger <sup>o</sup>, I. Michel-Behnke <sup>p</sup>, J.H. Nuernberg <sup>q</sup>, M. Peuster <sup>k</sup>, K. Walter <sup>r</sup>, P. Zartner <sup>s</sup>, F. Uhlemann <sup>t</sup>

<sup>a</sup> Deutsches Herzzentrum Berlin, Germany

<sup>b</sup> Medizinische Hochschule Hannover, Germany

<sup>c</sup> Universitätsklinik Bonn, Germany

<sup>d</sup> Herzzentrum Leipzig, Germany

<sup>e</sup> Universitätsklinik Freiburg, Germany

<sup>f</sup> Deutsches Herzzentrum München, Germany

<sup>g</sup> Universitätsklinik Köln, Germany

<sup>h</sup> Universitätsklinik Kiel, Germany

<sup>i</sup> Landes-Kinderklinik Linz, Germany

<sup>j</sup> Universitätsklinik Heidelberg, Germany

<sup>k</sup> Herz- und Diabeteszentrum Nordrhein-Westfalen, Germany

<sup>l</sup> Universitätsklinik Wien, Austria

<sup>m</sup> Universitätsklinik Erlangen, Germany

<sup>n</sup> Kinderspital Zürich, Switzerland

<sup>o</sup> Universitätsklinik Homburg, Germany

<sup>p</sup> Universitätsklinik Giessen, Germany

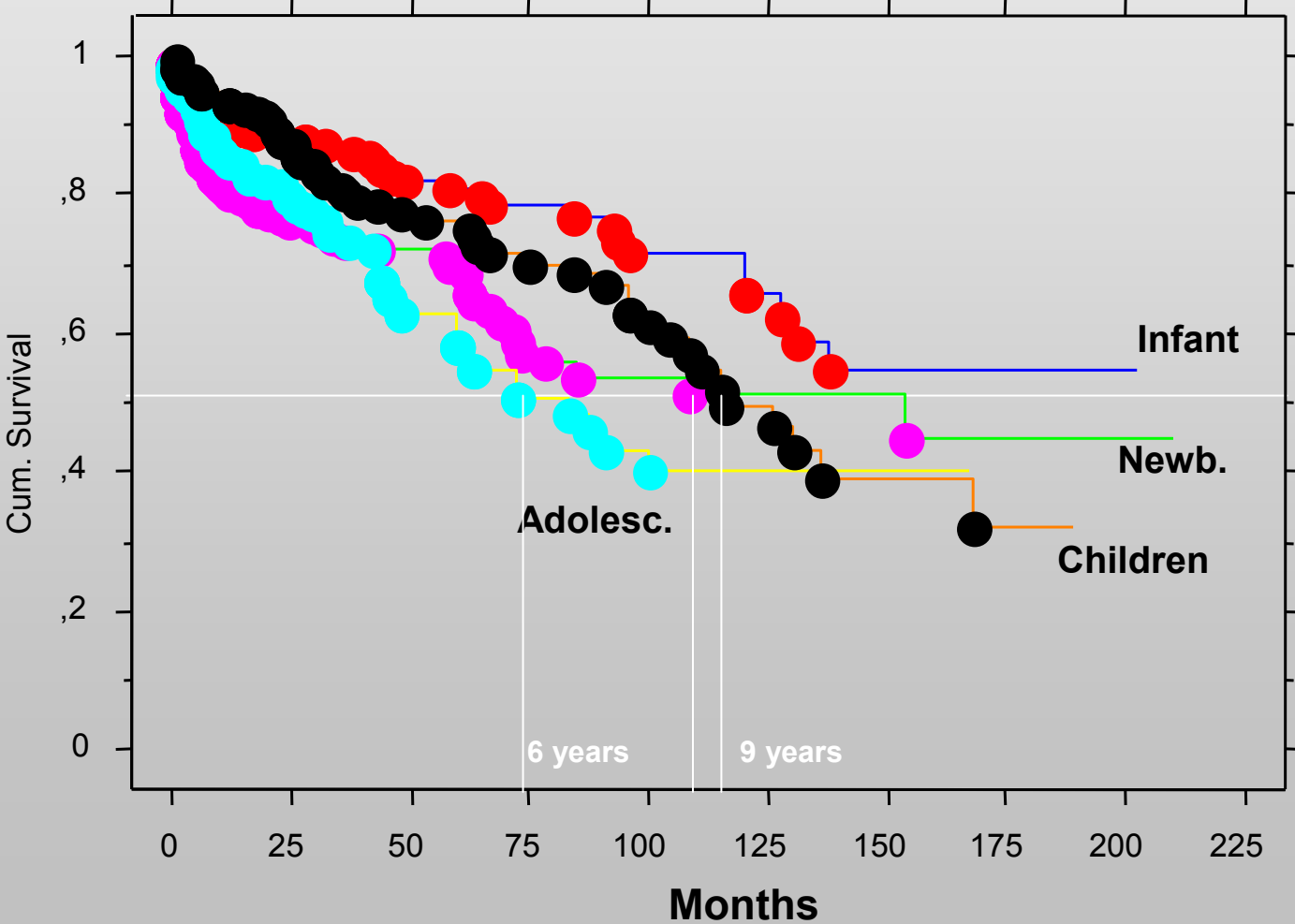
<sup>q</sup> Klinikum Links der Weser, Bremen, Germany

<sup>r</sup> Herzzentrum Duisburg, Germany

<sup>s</sup> Deutsches Kinderherzzentrum St. Augustin, Germany

<sup>t</sup> Olgahospital Stuttgart, Germany

# Kaplan-Meier curve for the event “operation”; patients divided by age





## 5) Therapy

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### 5.3) Therapy of valvular aortic stenosis beyond neonatal age

In these patients balloon valvuloplasty and commissurotomy are also competing possibilities.

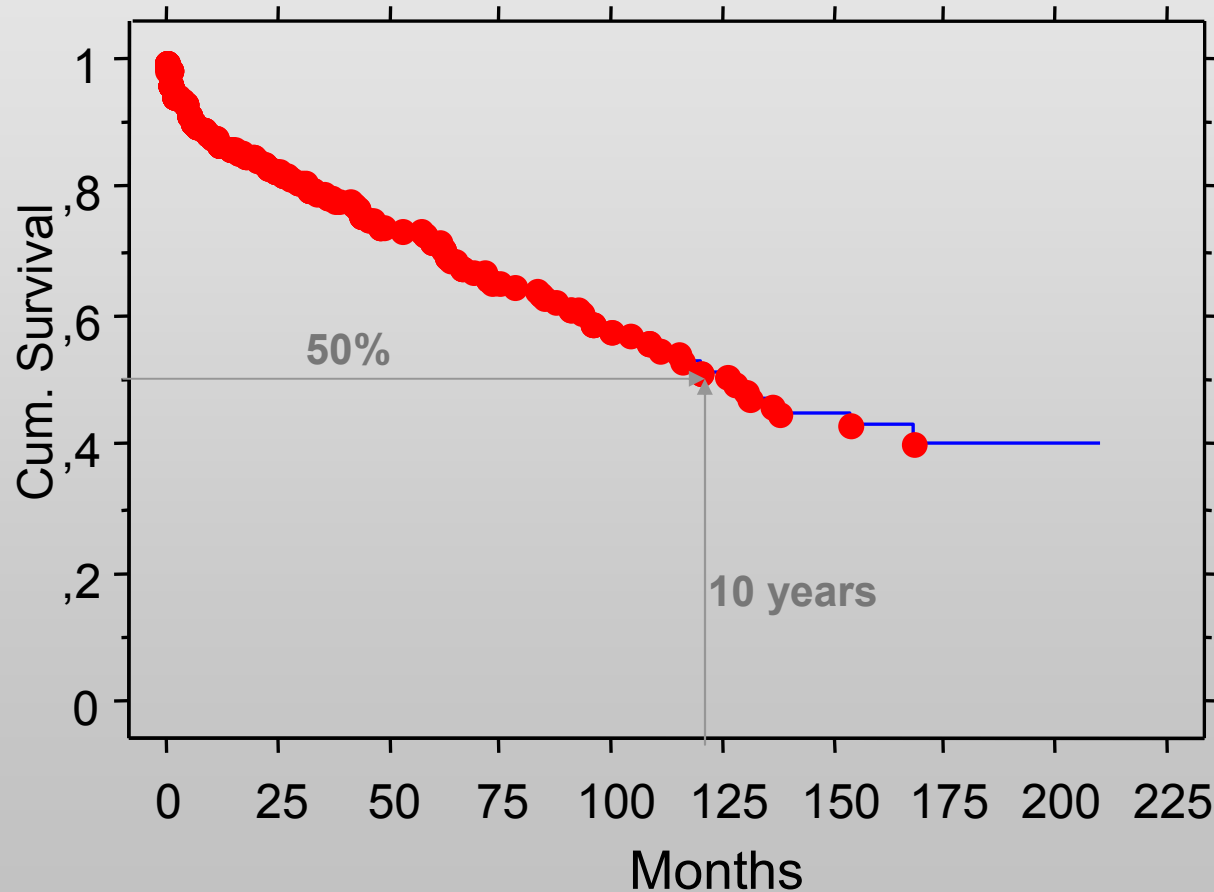
#### 5.3.1) Herz catheterization

Balloon valvuloplasty continues to show good results beyond neonatal age. It can make a valve operation unnecessary for many years and therefore reduce the total number of valve operations. In the ideal case a successful intervention can achieve such good valve function that the first operation can be delayed until the patient is an adult. However, with increasing age, additional aortic valve insufficiency gains more importance. This can develop spontaneously or as a residual effect of previous therapies (balloon dilatation or commissurotomy).

From grade II aortic sufficiency onwards balloon dilatation is no longer a promising option: there is a danger of higher grade insufficiency through the operation.

# Outcome after aortic balloon valvuloplasty in children

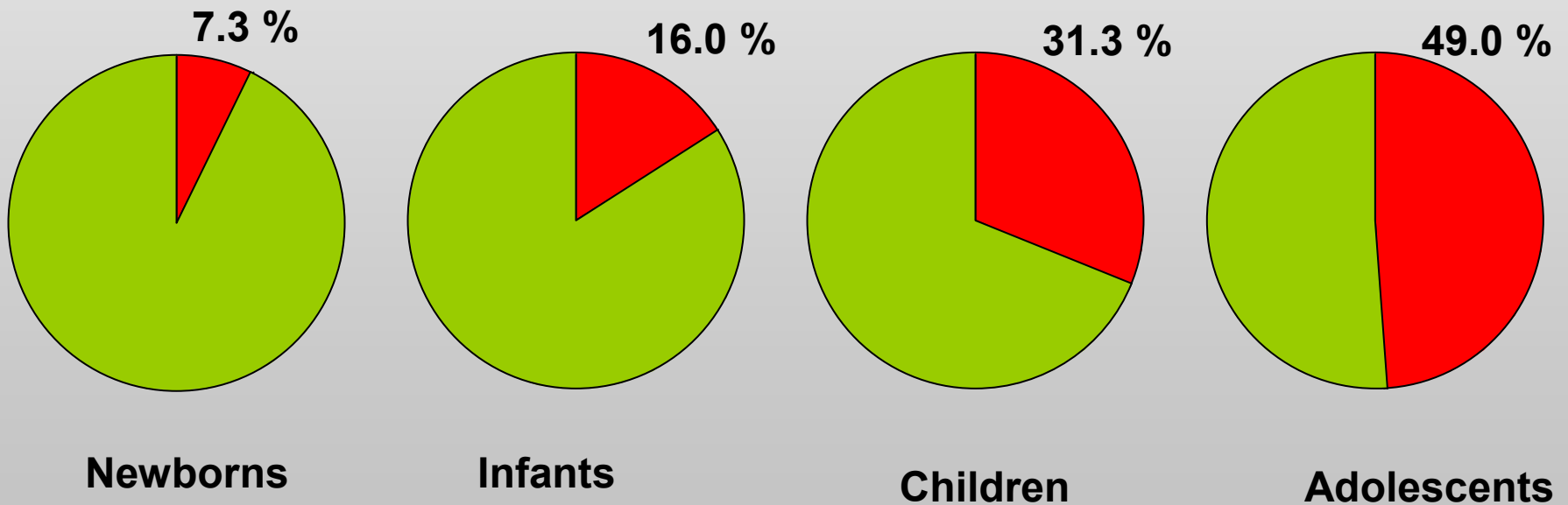
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**After 10 years 50% of the children are still free from operation**

# Aortic valve insufficiency before intervention

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## 5) Therapy

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### 5.3.1.1) Technique of catheter intervention

Normally retrograde access through the femoral artery is chosen. In newborns antegrade access through the umbilical vein or the femoral vein and an atrial septal defect is also possible.

The intervention should be begun with a balloon / valve relation of 0.8-0.9. The balloon diameter should not normally exceed that of the valve ring.

During the procedure, particularly in older children, functional cardiac arrest by rapid ventricular stimulation or adenosine may help to stabilize the balloon.

The risks attached to the intervention beyond newborn age are only slight.

# 5) Therapie

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## **5.3.2) Surgical procedures**

**5.3.2.1) Commissurotomy with and without aortic valve reconstruction**

**5.3.2.2) Ross operation**

**5.3.2.3) Mechanical valve replacement**

**5.3.2.4) Biological valve replacement**

## **5.3.3) Interventional aortic valve replacement**

Interventional valve replacement is not possible in children. At present it is recommended only in patients over 75 years of age due to the limited valve durability, the large access systems and the placement of a permanent stent. This kind of treatment is not to be expected for children for many years.

# 5) Therapy

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## 5.3.2) Surgical procedures

### 5.3.2.1) Commissurotomy with and without aortic valve reconstruction

Simple commissurotomy – like balloon dilatation – depends on the valve apparatus being well developed and severe valve insufficiency being absent. In some cases it is possible to treat the valve by commissurotomy and/or other repair procedures and at the same time to reduce or remove additional insufficiency. If the result of the reconstruction is seen intraoperatively to be unsatisfactory, the valve must be replaced.

# 5) Therapy

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## 5.3.2) Surgical procedures

### 5.3.2.2) Ross operation

In the Ross operation the aortic valve is replaced by the patient's own pulmonary valve (autograft) and a biological valve is implanted in pulmonary position. Anticoagulation is not necessary. The operation can be performed at any age and offers the advantage that a vital valve with growth potential is placed in aortic position. However, due to unphysiological pressure and dilatation of the genuine pulmonary valve, failure of the autograft is possible.

The disadvantage lies in the pulmonary valve replacement, which makes re-operations (valve exchange) necessary in the future. The optimal pulmonary valve prosthesis for the Ross operation has not yet been found. Even with decellularized biological prostheses, experience with valve replacement in children is limited and not very encouraging.

An increasing proportion of these defective pulmonary valves are being replaced by catheter intervention.

# 5) Therapy

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## 5.3.2) Surgical procedures

### 5.3.2.3) Mechanical valve replacement

Mechanical valve replacement offers the advantage that, in the ideal case, the valve life is unlimited. The disadvantage is the need for permanent oral anticoagulation. This is particularly suboptimal in children since traumatic bleeding complications are more likely due to their level of physical activity.

In addition, regular coagulation checks limit patients' quality of life.

In a later pregnancy the mechanical valve and the anticoagulation represent elevated risk factors. Therefore adapted anticoagulation management is necessary during pregnancy to reduce the risks for mother and child.

In childhood it must be taken into consideration that the valves do not grow. They should therefore not be implanted until somatic growth is complete and the anatomy of the LV outflow tract allows the implantation of a permanent valve.



## 5) Therapy

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### 5.3.2) Surgical procedures

#### 5.3.2.4) Biological valve replacement

The biological valve prosthesis may be a homograft or a xenograft. This is preferred when destructive aortic valve endocarditis is present. It also has the advantage that valves of different sizes may be stored (this applies at the moment only to homografts, whose availability is limited) and strict anticoagulation is not necessary. The disadvantage is the limited prosthesis durability. It is therefore suitable to bridge a limited time (several years) until definitive mechanical valve replacement is possible. This is advantageous in small children, in whom the valve ring does not yet allow mechanical valve replacement or in whom anticoagulation is to be avoided.

Further, it can be considered for women who wish to have children.

## 6) Follow-up

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Follow-up examinations should check the valve and ventricular function as well as the heart rhythm.

In addition, the procedures for endocarditis prophylaxis must be performed.

If the patient has a mechanical valve it is best if the coagulation checks can be performed by the parents or – in the case of adolescents – by the patient him- or herself.

## 6) Follow-up

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### 6.1) Valve function

After valve-preserving treatment (balloon valvuloplasty, surgical commissurotomy or aortic valve reconstruction) and after valve replacement with a biological valve (autograft, homograft, xenograft), echocardiography is necessary to check residual gradients and aortic valve insufficiency.

Following mechanical valve replacement, functional re-stenosis is to be expected as the patient grows, if growth-dependent size mismatch occurs. Further stenoses are possible through pannus formation or thromboses, which can restrict the movement of the prosthesis.

Insufficiency of a mechanical aortic valve is very rare but paravalvular leaks of hemodynamic relevance may occur. In some cases they can be treated by intervention; otherwise surgery is necessary.

After the Ross operation the function of the biological valve in pulmonary position must also be checked.

## 6) Follow-up

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### 6.2) Ventricular function

After treatment of the valve stenosis the LV function should normalize. However, it can remain impaired, even after successful restoration of valve function. This must be taken into account in follow-up examinations and may make supportive medication for heart failure necessary.

### 6.3) Rhythmus

In asymptomatic patients performance of long-term ECG and stress ECG examinations as a routine is not necessary.

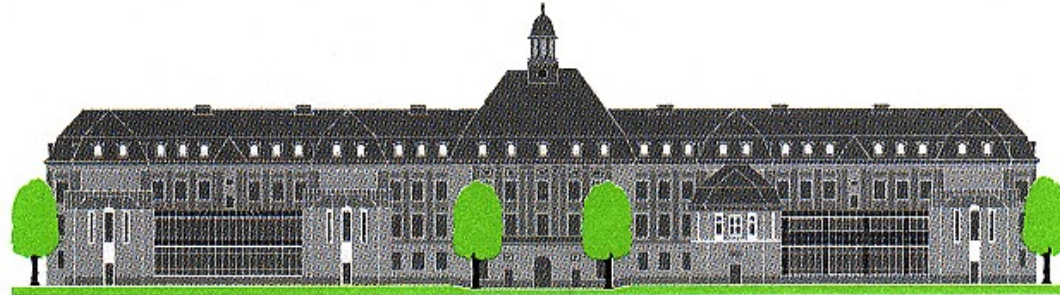
Following successful treatment of aortic valve stenosis, in rare cases ventricular rhythm disturbances may occur. Long-term ECG and stress ECG are then recommended.

## 7) Prevention

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There is no means of preventing isolated congenital aortic valve stenosis.

The opportunity for genetic counselling and prenatal diagnostic procedures, including fetal echocardiography, should be offered.



**DEUTSCHES HERZZENTRUM BERLIN**  
STIFTUNG DES BÜRGERLICHEN RECHTS

## **Guidelines: Congenital Aortic Stenosis**

**PD Dr. med. Peter Ewert**

Senior Physician, Department of Congenital Heart Disease / Pediatric Cardiology,  
Deutsches Herzzentrum Berlin