

# Cardiology in the Young

Special Supplement of *Cardiology in the Young*:  
The German Society of Paediatric  
Cardiology (DGPK)  
Guidelines for the Management  
of Congenital Heart Diseases  
in Childhood and Adolescence



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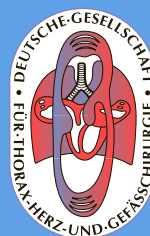
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*In collaboration with:*



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### The German Society of Paediatric Cardiology (DGPK) Guidelines for the Management of Congenital Heart Diseases in Childhood and Adolescence

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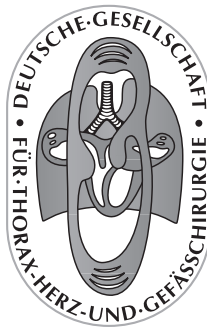
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**General remarks**

The German Society of Paediatric Cardiology (DGPK) was founded in 1973 as the first subspecialty of Pediatrics in Germany. The DGPK is a non-commercial non-profit medical organization. Aims are to promote scientific research, diagnosis and therapy as well as prevention of congenital and acquired cardiovascular malformations and diseases in childhood and adolescence. One major task of the society is education and training of physicians working in the field of paediatric cardiology. For this purpose, guidelines constitute an important and effective instrument for assurance of a high level of quality and continuous medical education.

The DGPK has continuously developed guidelines for specific entities since 1995. In general, guidelines in Germany have to be developed in accordance with the rules of the German Association of the Scientific Medical Societies (AWMF).

*German Association of the Medical Professional Societies (AWMF)*

The German Association of the Scientific Medical Societies (“Arbeitsgemeinschaft der Wissenschaftlichen Medizinischen Fachgesellschaften e.V.” - AWMF), founded in 1962, is the head organization of more than 150 German medical societies.

AWMF advises the government of the Federal Republic of Germany as well as the governments of the states in all areas of scientific medicine and medical research. AWMF is the representative of Germany in the “Council for International Organizations of Medical Sciences (CIOMS)” of World Health Organization (WHO) in Geneva (Switzerland) as well a member of the “Guidelines International Network (GIN)”.

An important task of AWMF is the coordination of medical guidelines. AWMF guidance is designed to provide a tool for the scientific medical societies to create and publish up-to-date and high-quality guidelines in the AWMF Guideline Register (<http://www.awmf.org/leitlinien/awmf-regelwerk.html>).

AWMF guidelines are defined as “systematically developed statements reflecting the current state of knowledge aiming to support doctors and patients in making decisions concerning appropriate care for specific health problems. Their primary objective is to

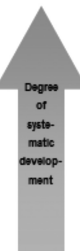
improve medical care by disseminating current knowledge”.

Guidelines differ from other sources of “processed” knowledge (e.g., text books or systematic reviews) as they express clear consensus- and/or evidence-based recommendations.

*Classification of AWMF guidelines*

The AWMF classification ranges from the lowest level, i.e. a guideline based on expert recommendations S1, to intermediate level S2k or S2e guidelines up to the highest level, i.e. evidence- and consensus-based S3 guidelines (see below).

**S-classification**  
according to the AWMF Guidance Manual and Rules

<b>S3</b>	<b>Evidence- and consensus-based guideline</b>	Representative committee, Systematic review and Synthesis of the evidence, structured consensus development	 Degree of systematic development
<b>S2e</b>	<b>Evidence-based guideline</b>	Systematic review and Synthesis of the evidence	
<b>S2k</b>	<b>Consensus-based guideline</b>	Representative committee, structured consensus development	
<b>S1</b>	<b>Recommendations by expert groups</b>	Consensus development in an informal procedure	

Every class represents a specific methodological concept of generating guidelines. Furthermore, the class indicates the level of evidence available or evaluated.

The level of consensus-based S2k class was chosen by the DGPK guidelines, since there are no sufficient evidence-based publications or prospective randomized trials available in paediatric cardiology.

The following prerequisites are mandatory to generate a S2k-guideline:

- All relevant societies, associations or organizations which are involved in a given topic are represented in the guideline committee.
- Scientifically established formal consensus methods like the Nominal Group Process, the Structured Consensus Conference and the Delphi Technique are used. Structured consensus development ensures that relevant key questions and recommendations are discussed without restrictions.
- A neutral moderator is in charge to supervise discussions and subsequent voting on recommendations.



- Classes of recommendations and levels of evidence are not stated in S2k guidelines due to a lack of appropriate evidence-based studies or publications. Therefore, only consensus based recommendations can be provided.

### Guidelines of the German Society of Paediatric Cardiology (DGPK)

For this purpose the executive board of the DGPK has set up a guideline committee in order to establish and up-date the guidelines. Members of the guideline committee are proposed by the DGPK executive board and acknowledged during the annual general assembly of the DGPK.

The guideline committee is filled with representatives of the following organizations:

- German Society of Paediatric Cardiology (DGPK)
- German Society of Cardiac and Cardiovascular Surgery (DGTHG)
- Working group of Paediatric Cardiologists working in private practice (ANKK)
- Working group of Paediatric Cardiologists working in general paediatric hospitals (AAPK)
- German Heart Foundation (DHS)
- German umbrella organization for congenital heart diseases (parents organization, BVHK)

All guidelines were approved by the executive board of the DGPK as well as the German Society for Cardiac and Cardiovascular Surgery (DGTHG) for guidelines when surgical treatment was involved. Furthermore guidelines were acknowledged by the executive board of the German Society for Child and Adolescent Medicine (DGKJ) and - if appropriate - by other societies such as the German Society of Cardiology (DGK).

All guidelines were established without private or industry driven sponsoring. All expenses were provided by the DGPK and the German Heart Foundation (DHS). We are very grateful for the generous financial support of the DHS.

All members of the guideline committee disclosed their written conflict of interests according to the policy of the AWMF. Work of all members was on a voluntary basis and without any financial reward.

#### *The first DGPK Guidelines in German language*

The first edition of the DGPK guidelines were published in 1998. Thereafter guidelines were continuously revised and republished as a hard copy in 2007 and recently in 2016 as S2k guidelines as hard copy and

e-book by Elsevier Publishing Company (ISBN print 978-3-437-22391-4 and ISBN e-book 978-3-437-29919-3).

Recently revised guidelines are accessible on the homepage of the DGPK ([www.kinderkardiologie.org/leitlinien](http://www.kinderkardiologie.org/leitlinien)) and on the homepage of the German Association of the Medical Professional Societies ([www.awmf.org/leitlinien](http://www.awmf.org/leitlinien)).

#### *Short version of the DGPK Guidelines in English*

All DGPK guidelines were reedited as a shortened version in English containing key messages and recommendations of the respective long-version German guideline.

All long-version guidelines as the basis of the present shortened versions had been established at the S2k level of the AWMF classification (see above). Therefore, as in the long-versions, only recommendations but no information about classes of recommendations and levels of evidence are provided in these shortened versions.

Each particular English guideline was created by the authors of the respective long-version guideline. All guidelines were subsequently discussed and approved by the guideline committee. All authors provided a statement on their conflict of interests concerning their respective guideline. All members of the guideline committee are listed below together with their declaration concerning conflict of interest.

For the purpose of brevity only a limited number of key references is given. The long list of references is accessible in the original German version by the link which is indicated at the end of each guideline.

The shortened version of guidelines is intended to support physicians and all other health care providers dealing with cardiovascular diseases in childhood and adolescence by providing a quick overview on key diagnostic and therapeutic issues in specific diseases. These guidelines are not intended to substitute textbooks for pediatric cardiology. They may provide additional support for physicians in their daily clinical routine work.

The final decision in any particular patient must be established by the physician(s) in charge. Finally, the physician is responsible to observe any rules and regulations applicable to drugs, devices and procedures.

We hope that these guidelines will be of support and guidance for all health care workers who are involved in the care of children and adolescents with diseases of the cardiovascular system.

Any comments and/or proposals for improvements will be appreciated.

# Members of the Guideline Committee

## DGPK German Society of Paediatric Cardiology

**Chairman: Prof. Dr. med. Jochen Weil**

Organisation: Department of Paediatric Cardiology and Congenital Heart Disease  
German Heart Centre Munich, München

Conflicts of interest: Lectures for Actelion and GlaxoSmithKline

Representative: Prof. Dr. med. Ingo Dähnert

Organisation: Department of Paediatric Cardiology and Congenital Heart Disease  
University Heart Center Leipzig, Leipzig

Conflicts of interest: Cooperations with industry (since 2013): Lectures Actelion, Gore, Boston Scientific;  
Proctor: Metronic, Occlutech, St. Jude; Consultant: Acoredis, Boston Scientific; Clinical trials:  
Occlutech, Osypka, Otsuka, Pfizer, Lifetec

Representative: Prof. Dr. med. Harald Bertram

Organisation: Department of Paediatric Cardiology and Congenital Heart Disease  
Hannover Medical School, Hannover

Conflicts of interest: None

Representative: Prof. Dr. med. Sven Dittrich

Organisation: Department of Paediatric Cardiology and Congenital Heart Disease  
University Erlangen, Erlangen

Conflicts of interest: Honoraria for lectures from Berlin Heart and travel compensations for industrial meetings from Medtronic and Siemens. His institution (Dept. Pediatric Cardiology University Hospital Erlangen) received compensation for contributions to clinical trials from Novartis, Bayer, Servier and Ozuka. The institution received unrestricted scientific grants from Siemens and Materialize and funding for scientific meetings from AbbVie, Actelion, Orion Pharma and St. Jude

Representative: Prof. Dr. med. Matthias Gorenflo

Organisation: Department of Paediatric Cardiology and Congenital Heart Disease  
University Heidelberg, Heidelberg

Conflicts of interest: Lecture Fees: Actelion; Bayer Schering; GlaxoSmithKline

Representative: Prof. Dr. Nikolaus Haas

Organisation: Department of Paediatric Cardiology and Congenital Heart Disease  
University Munich, Campus Großhadern, München

Conflicts of interest: None

Representative: Prof. Dr. med. Alfred Hager

Organisation: Department of Paediatric Cardiology and Congenital Heart Disease  
German Heart Centre Munich, München

Conflicts of interest: Encysive GmbH, Pfizer, Actelion Abbott, Medtronic, Schiller Medizintechnik GmbH, GlaxoSmithKline, AOP Orphan Pharmaceuticals AG, OMT. He received Honoraria for writing informational material from Actelion. He received travel compensations from Braun, Guidant, Arrows, Medtronic, Actelion, GlaxoSmithKline, Pfizer, Lilly, AOP Orphan Pharmaceuticals AG. He is shareholder of Johnson & Johnson, Gilead, Merck Sharp & Dohme Inc., Pfizer, Medtronic, Roche, and many other Biotech companies. His institution received compensations for contribution to company driven clinical trials from Actelion, Medtronic, Edwards, Occlutech, Novartis, Lilly and other companies. His institution received unrestricted scientific grants for investigator initiated trials from Pfizer, GlaxoSmithKline, Abbott, Actelion, Medtronic and other companies.

Representative: Prof. Dr. med. Rainer Kozlik-Feldmann  
Organisation: Department of Paediatric Cardiology and Congenital Heart Disease  
University Heart Center Hamburg, Hamburg  
Conflicts of interest: 2015 has been referent for Actelion and today proctor for pfm Medical AG

Representative: Prof. Dr. med. Angelika Lindinger  
Organisation: Previous Department of Paediatric Cardiology and Congenital Heart Disease  
University Saarland, Homburg  
Conflicts of interest: None

Representative: Prof. Dr. med. Thomas Paul  
Organisation: Department of Paediatric Cardiology and Congenital Heart Disease  
University Göttingen, Göttingen  
Conflicts of interest: None

Representative: Prof. Dr. med. Carsten Rickers  
Organisation: Department of Paediatric Cardiology and Congenital Heart Disease  
University Kiel, Kiel  
Conflicts of interest: None

#### **DGTHG German Society for Thoracic and Cardiovascular Surgery**

Representative: Prof. Dr. med. Robert Cesnjevar  
Organisation: Department of Paediatric Cardiac Surgery  
University Erlangen, Erlangen  
Conflicts of interest: None

Representative: Prof. Dr. med. Oliver Dewald  
Organisation: Department of Cardiac Surgery  
University Bonn, Bonn  
Conflicts of interest: Medtronic (educational support), Actelion (educational support)

Representative: Prof. Dr. Peter Murin  
Organisation: Department of Paediatric Cardiac Surgery  
German Heart Centre Berlin, Berlin  
Conflicts of interest: None

#### **BVHK German umbrella organization for Congenital Heart Disease (parents organization)**

Representative, Dr.med. Raphael Dorka  
Organisation: Department of Paediatrics  
Hospital, Landau  
Conflicts of interest: None

Representative: Prof. Dr. Phil. Elisabeth Sticker  
Organisation: Department of Psychology  
University Köln, Köln  
Conflicts of interest: None

#### **ANKK Working Group of Paediatric Cardiologists working in private praxis**

Representative: Dr. med. Karl-Robert Schirmer  
Organisation: Paediatric Cardiologist, Hamburg  
Conflicts of interest: None

Representative: Dr. med. Marc Schlez  
Organisation: Paediatric Cardiologist, Neustadt/Weinstraße  
Conflicts of interest: None

#### **AAPK Working Group of Paediatric Cardiologists working in paediatric hospitals**

Representative: Dr. med. Liane Kändler  
Organisation: Department of Paediatrics  
Hospital Wittenberg, Wittenberg  
Conflicts of interest: None

Representative: Dr. med. Stefan Stuhmann  
Organisation: Department of Paediatrics  
Hospital Offenburg, Offenburg  
Conflicts of interest: None

#### **DHS German Heart Foundation**

Representative: Kai Ruenbrink  
Organisation: Deutsche Herzstiftung (German Heart Foundation, DHS)  
Frankfurt  
Conflicts of interest: None

#### **Moderator**

Representative: Prof. em. Prof. h.c. Dr.med. Achim A. Schmaltz  
Organisation: Managing Director of the German Society of Paediatric Cardiology (DGPK)  
Düsseldorf  
Conflicts of interest: None

# Association for European Paediatric and Congenital Cardiology (AEPC)



**A**EPC WAS FOUNDED IN LYON IN 1963 (as Association Européenne pour la Cardiologie Pédiatrique) and subsequently has created a network of specialists who are committed to the practice and advancement of Congenital Cardiology and closely related fields. *Cardiology in the Young* is the official journal of the AEPC. AEPC offers a free subscription of 'Cardiology in the Young' (CitY) as part of the annual membership fee. The membership also offers several other benefits.

The overall membership of the Association currently stands at 1300 paediatric cardiologists and other specialists working in the field of paediatric cardiology and its related disciplines. As far as we are aware, the AEPC is the largest democratically administered global association in the field of paediatric and congenital cardiology. Members of the AEPC originate from virtually all countries in Europe and it is encouraging that there are now increasing numbers of members from all the continents. New members are very welcome as they bring with them new ideas and innovations.

AEPC and its Working Groups aim to enhance collaboration amongst members for scientific research, promoting training, professional development and to maintain high standards of professional practice. The work on behalf of children and patients with congenital heart diseases is global and the AEPC has made itself a global organisation by its close collaboration with other international organizations. These include the European Society of Cardiology;

the Japanese Society of Paediatric Cardiology and Cardiac Surgery (JSPCCS); the European Association for Cardio-Thoracic Surgery (EACTS) and; the Adult Congenital and Pediatric Cardiology Section of the American College of Cardiology (ACC); the Asia Pacific Pediatric Cardiac Society (APPCS); the European Heart Surgeons Association (ECHSA), the World Congress of Paediatric Cardiology and Cardiac Surgery and several others.

## Working groups

Several activities of AEPC are organized by the Working Groups. The Working Groups represent different subspecialties and specific areas of paediatric and congenital cardiology. The Association now has 13 Working Groups, to bring together workers with similar interests in order to facilitate research and collaboration and to organise teaching and training.

## Annual meetings

An Annual Meeting and an Update-On Course are organised by the AEPC, usually in the third week of May in collaboration with one of the member countries. The AEPC organizes 2–3 Teaching courses for trainees in Paediatric Cardiology each year. Additional symposia and courses are usually a part of the annual meetings.

# The International Society for Nomenclature of Paediatric and Congenital Heart Disease



**T**HE INTERNATIONAL SOCIETY FOR NOMENCLATURE of Paediatric and Congenital Heart Disease (ISNPCHD) is an established not-for-profit organization, incorporated in Canada, with the collective mission to identify, standardize, and maintain an international system of nomenclature, the International Paediatric and Congenital Cardiac Code (IPCCC), to enhance global communication and facilitate patient care, research, and training in paediatric and congenital heart care across disciplines. In other words, the IPCCC provides a common language and terminology, inclusive of definitions and imaging, which covers the entire field of diagnostic and procedural terms for paediatric and congenital heart care. The IPCCC is the product of the cross-mapping work by the ISNPCHD Nomenclature Working Group during the decade following the publication in 2000 of two similar and complementary nomenclature systems, namely the International Congenital Heart Surgery Nomenclature and Database Project under the auspices of the European Association for Cardio-Thoracic Surgery and Society of Thoracic Surgeons, and the European Paediatric Cardiac Code under the auspices of the Association for European Paediatric Cardiology. The IPCCC is owned by the ISNPCHD but is digitally published for free download for private use ([www.ipccc.net](http://www.ipccc.net)) in these two Societal versions, both with a Long List of over 10,000 terms with qualifiers, and a Short List of up to 1000 terms for use in databases when comparing institutional outcomes, both nationally and internationally. The Long Lists of the IPCCC comprehensively cover the field of paediatric and congenital cardiac care, including diagnoses of congenital and related acquired pathology, comorbid conditions, transcatheter and operative procedures, and a full list of postprocedural complications. The Short Lists are used within databases across the world with over 500,000 registered patients. The ISNPCHD encourages the commercial use of the

IPCCC by requiring and providing free-of-charge license agreements for its use, to ensure that the IPCCC remains unaltered by parties other than the ISNPCHD.

Since 2007 the ISNPCHD has been operating through three working groups:

- The Nomenclature Working Group, which continues to maintain, develop, expand, update, and preserve the IPCCC;
- The Definitions Working Group, which is engaged in writing definitions for the terms in the IPCCC. More recently this initiative has focused on the terms provided by the ISNPCHD at the behest of the World Health Organization (WHO) for the 11<sup>th</sup> revision of the International Classification of Diseases;
- The Archiving Working Group, which is engaged in linking images and videos to the IPCCC, including cardiac morphologic specimens, echocardiography, angiography, computerized axial tomography, magnetic resonance imaging, intraoperative photographs and intraoperative videos.

The ISNPCHD, through the IPCCC, enables institutions from around the world to seamlessly communicate with each other, comparing and then improving outcomes and the quality of care that is given to children, young people, and adults born with malformed hearts. This common language enables institutions to learn from those hospitals performing best at a global level, as well as facilitating research projects, such as comparing the longer term quality of life and complications in those who have required operative and transcatheter interventions. In addition, the nomenclature with corresponding definitions and matching imaging, enhances teaching of this specialty to the next generation of clinicians dedicated to pediatric and congenital cardiac care, both in the developed and developing world.