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

Supplement Editor: Jochen Weil, MD, PhD
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Professor for Paediatric Cardiology and Congenital Heart Disease
Consultant at the German Heart Center, Munich (Germany)

In collaboration with:

German Society for Thoracic and Cardiovascular Surgery (DGTHG)
German Heart Foundation

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(Short versions)

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Deutsche Herzstiftung

German Heart Foundation
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Jochen Weil

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Introduction

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).

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 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.

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 healthcare 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.

**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 healthcare 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.

**The first DGPK Guidelines in German language**


Recently revised guidelines are accessible on the homepage of the DGPK (www.kinderkardiologie.org/leitlinien) and on the homepage of the German Association of the Medical Professional Societies (www.awmf.org/leitlinien).
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
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Representative: Prof. Dr. med. Sven Dittrich
Organisation: Department of Paediatric Cardiology and Congenital Heart Disease
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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 Otsuka. The
institute received unrestricted scientific grants from Siemens and Materialize and funding for scientific
meetings from AbbVie, Actelion, Orion Pharma and St. Jude

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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. institutional DHM. His institution received compensations for contribution to company
driven clinical trials from Actelion, Medtronic, Edwards, Occlutec, Novartis, Lilly and other companies.
His institution received unrestricted scientific grants for investigator initiated trials from Pfizer,
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Conflicts of interest: 2015 has been referent for Actelion and today proctor for pfm Medical AG

Representative: Prof. Dr. med. Angelika Lindinger
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Representative: Prof. Dr. med. Thomas Paul
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Conflicts of interest: None

Representative: Prof. Dr. med. Carsten Rickers
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Representative: Prof. Dr. med. Oliver Dewald
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BVHK German umbrella organization for Congenital Heart Disease (parents organization)
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ANKK Working Group of Paediatric Cardiologists working in private praxis
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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
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Conflicts of interest: None

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

DHS German Heart Foundation

Representative: Kai Rüenbrink
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
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’ (CiY) 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

The 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) 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 11th 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.