

SPECIAL LECTURE 1

Competence Network for Congenital Heart Defects (Kompetenznetz Angeborene Herzfehler)

[Objectives]

The Competence Network for Congenital Heart Defects is national research project funded by the Federal Ministry of Education and Research (BMBF) since 2003. It combines basic interdisciplinary medical research into congenital heart defects with clinical research and medical care. Doctors, scientists, parent associations and self-help groups work together in the network. The aim is to integrate the latest scientific findings more rapidly in medical practice and to guarantee better care for patients with congenital heart defects.

[Focus of activities] Congenital heart defects are the most frequent congenital illness. Each year in Germany, about 6,000 children are born with such defects. Owing to the progress made in pediatric cardiology, heart surgery, and anesthesia, over 90% of patients now reach adulthood. The majority – currently about 300,000 young children, adolescents, and adults – remain chronically ill for life.

The network focuses on

- setting up an efficient research structure,
- ensuring the quality of research and care,
- informing doctors, patients, and the public about congenital heart defects.

Multi-centric clinical research

The number of patients in individual research centers is small. A networked research structure on congenital heart defects is therefore the basis for advances in the treatment of this syndrome.

This is the only way to obtain data of a sufficiently large number of patients to allow the results of studies to qualify as representative according to international peer review standards. The network's multi-centric studies focus on new diagnostic and treatment methods, the development of guidelines and treatment standards, and the epidemiology of congenital heart defects.

National register for congenital heart defects

Patient with congenital heart defects have an excellent chance of survival, so their numbers are continuing to rise. But there is not much reliable data available on these patients, anywhere in the world.

There is a serious lack of data on prevalence, morbidity, mortality, life expectancy and quality of life that takes into account the latest diagnostic and treatment options.

The National register for congenital hearts defects was set up to fill this gap. Ultimately it is hoped to obtain records for all patients in the country, providing a basis for epidemiological studies on congenital heart defects.

[Projects]

The main focus of the multi-centric studies conducted by the Competence Network for Congenital Heart Defects are research projects concerning the right ventricle, which tends to be affected most seriously in such

cases. The aim is to establish standards for non-invasive examination procedures (echocardiography, magnetic resonance imaging) and a set of treatment guidelines. Epidemiological studies – on the prevalence of congenital heart defects, their long-term progression, the patients’ quality of life and treatment options – are conducted for input to the national register. The data collected during these studies are compiled in an uniform record format, enabling all users to obtain the maximum added value from them.

[Innovation highlights]

European research alliance on the genetics of congenital heart defects

As part of the major European research project on “Heart Failure and Cardiac Repair”, the teams participating in the national register are conducting research into the genetic factors that give rise to congenital heart defects. The register plays an important role as a phenotype database for the collection and supply of clinical data concerning the diagnosis, prognosis, life expectancy, quality of life and treatment options of the patients under investigation.

The research consortium is funded under the Sixth EU Framework Programme and comprises 27 leading representatives of this field from Sweden, the UK, Germany, the Netherlands, France, Spain, Italy and Switzerland. Various study groups are investigating such topics as the genetics of heart muscle formation or cell interaction and renewal. In related projects, techniques are being developed to facilitate and accelerate the repair of damaged heart-muscle cells.

Electronic patient files

The Competence network stores a lifelong record of the medical data on patients with congenital heart defects in its research database and the epidemiological register. The resulting electronic patient files can be used as a prospective healthcare resource under the strict application of the relevant data protection requirements.

The stored data can be linked with those on the electronic health insurance card, where they can be of immense benefit in the treatment of patients with chronic heart disease.

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SPECIAL LECTURE 2

Quality of life and psychosocial aspects: First results from the National Register for congenital heart defects

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Objectives: The medical and surgical progress of the last decades has resulted in dramatic improvements in terms of increased life expectancy and thus gave rise to a new population of chronically ill patients facing new psychosocial challenges. Where treatment but complete cure is not always available the measurement of quality of life gains an essential value for the long-term surveillance.

Methods: A questionnaire was sent to all adult patients enrolled in the National Register (n=4397). The SF-36, an international validated questionnaire with good psychometric quality, is used for the measurement of the health related quality of life. The data from the German National Health Survey allows to draw a comparison with a normative sample of the German population.

Results: On the basis of the data from the National Register a comprehensive overview of the psychosocial aspects affecting the adult life of persons with congenital heart defects can be given. It is determined if and - if yes - to what extent adults with congenital heart disease have an impaired quality of life compared to a healthy population. Potential social and psychological factors that have an influence on the quality of life and the emotional functioning of these patients are being analyzed. The influence of other factors such as cardiac diagnosis, therapy outcome or physical resilience on the subjective quality of life can also be quantified.

Conclusions: Due to increasing numbers of adults with congenital heart defects there is an urgent need for information on the specific challenges experienced by our patients in their daily life. This study's results by now do suggest that patients with CHD are to a good extent successfully engaging in full adult responsibilities and roles but that they also have to face specific psychosocial experiences. This data will help to identify factors important for a successful emotional and social integration of the diagnosis CHD into a satisfying adult life. As will be shown the association between the empirically measurable somatic findings and the subjective quality of life is weak; therefore, a more multidisciplinary approach including for example a special psychosocial therapy becomes crucial.

SPECIAL LECTURE 5

Arrhythmia in complex univentricular heart after Fontan operation

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Because of recent advance of surgical results, many children with complex congenital heart disease have survived after corrective or palliative repair. However, tachyarrhythmia is recognized to be an important source of early and late morbidity and mortality especially in post-Fontan patients. Atrial tachycardia had been reported to occur in 11~ 42% of patients following Fontan operation. Atriopulmonary connection, older age, sinus node dysfunction, and atrial thrombus were known to increase the risk of late atrial tachycardia. Atrial flutter and atrial fibrillation have been known to be major tachycardias following Fontan type palliation. Various treatment modalities such as antiarrhythmic medications, pacemaker implantation, transcatheter ablation, and Fontan conversion with arrhythmia surgery have been tried successfully. In Fontan patient, the reentry circuits serving atrial flutter are usually complicated with multiple circuits. Transcatheter ablation for atrial flutter in Fontan patients is challenging condition in both mapping and making definitive ablation lesion, so acute success rate was reported to be 50-75% and recurrence rate in 1 year was 40-50%. In our institution, sixteen cases have undergone Fontan conversion surgery because of atrial arrhythmia or atrial thrombosis. Two of them had protein losing enteropathy. Fourteen out of sixteen showed symptomatic improvement after Fontan conversion and arrhythmia surgery. Recently I and my colleagues reported the coexistence of two distinct AV nodes in complex congenital heart disease and its incidence and relation to arrhythmias. From 2001 to 2003, we performed an electrophysiologic study upon 52 consecutive patients who had undergone cardiac catheterization after Fontan completion. Atrial pacing was performed at 3 or more different atrial sites. In 10/52 patients, two different QRS complexes were recorded at different pacing sites, suggesting twin AV node (9/20 in right atrial isomerism, 1/8 AV discordance, 0/24 other complex anomalies). AV reciprocating tachycardia (AVRT), presumably involving two AV nodes and a connecting sling, was induced in 6 of 10 patients who had twin AV node (4/6 used posterior AV node as an antegrade limb, 2/6 used an anterior AV node as an antegrade limb). Heterotaxy syndrome ($p < 0.001$) and complete atrioventricular septal defect ($p = 0.002$) were found to be risk factors for twin AV node. Junctional tachycardia (JT; $HR > 150/\text{min}$) with either VA dissociation (7/9) or second degree VA block (2/9) were induced by pacing or isoproterenol infusion in 9/52 patients. Junctional tachycardia induction was associated with a twin AV node ($p = 0.04$), or a history of early postoperative junctional ectopic tachycardia ($p = 0.02$). A complicated AV node conduction system such as twin AV node was frequent in heterotaxy syndrome. Both AVRT, and JT with VA block may be important causes of tachyarrhythmia in this patient group.