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## Guidelines

## Recommendations to organize care for adults with congenital heart disease in the Czech Republic



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<https://doi.org/10.1016/j.crvasa.2018.03.009>

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## ARTICLE INFO

## Article history:

Received 13 February 2018

Accepted 15 March 2018

Available online 13 April 2018

## Keywords:

adult congenital heart disease

ACHD

grown-up congenital heart disease

GUCH

organization of care

specialized centre

## ABSTRACT

This paper was formed by the Expert committee for congenital heart disease (CHD) in adults – a division of Czech Society of Cardiology (ČKS). It was designed as an appendix to *National cardiovascular programme ČKS* created in 2013 and was based on *Recommendations for organization of care for adults with congenital heart disease and for training in the subspecialty of 'Grown-up Congenital Heart Disease' in Europe: a position paper of the Working Group on Grown-up Congenital Heart Disease of the European Society of Cardiology* created in 2014 [1].

Aims of this paper are: To optimize medical care in all its aspects for adults with CHD in the Czech Republic, to facilitate easy transition between paediatric and adult medical care, to enable research in the field in order to create evidence based care, to support training of regional cardiologists and other specialists who are involved in monitoring adult patients with CHD, to help with communication with national institutions, to provide information for other medical workers and patients, to consolidate resources.

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## Introduction

Incidence of CHD is 6–8 per 1000 newborns. Progress of paediatric cardiology and cardiac surgery dating back to 8th decade of previous century led over time to significant increase in long-term survival of patients with CHD. Today about 90% of patients with CHD grow into adulthood [2]. The estimated prevalence of all CHDs is 12/1000 in children and 5/1000 in adults, the estimated prevalence of critical/severe CHDs is 1.45/1000 in children and 0.38/1000 in adults [3], while each year the absolute number of adults with CHD increases by 5% [4]. Total number of adults with CHD in the European Union is estimated around 2.3 million (for comparison total number of adults with ischaemic heart disease is 24 million) [5].

If we extrapolate above mentioned data, we should end up with around 53,000 adults with CHD (including atrial septal defects and bicuspid aortic valve) in the Czech Republic, 4000 of these would have a severe CHD. At the moment three complex centres specializing in CHD in the Czech Republic register 9000 adult patients diagnosed with CHD. Frequency of hospitalizations of adult patients with CHD has increased by 81.5% between 2003 and 2010 and mean age at time of hospitalization has increased from 53.5 years to 57.5 [6]. This increase of hospitalizations of adults with CHD has even overcome number of hospitalizations of children with CHD [7].

Due to significant decrease in early mortality in patients with CHD the modern medical care now encounters and has to deal with long-term complications of CHD such as chronic heart failure, high risk of infective endocarditis, higher occurrence of arrhythmias, pulmonary hypertension, valve

defects and complications during pregnancy. Wide range of CHD can be categorized as a rare disease (occurring in less than 5/10,000 people) [8]. Population of adult patients with CHD is and will always be relatively small but at the same time it is and will be very heterogeneous in spectrum of CHD and their particular complications and subsequent management. These guidelines in combination with super-specialized multi-disciplinary medical care are in accord with *National strategy for rare diseases for years 2010 to 2020*. This means that implementation of these guidelines into medical care should increase efficiency of diagnostic and treatment process of rare diseases (in our case CHD) and should provide all patients with rare disease with indicated and high-quality medical care and help them with social incorporation [9].

## Organization of care

System of medical care for adults with CHD in Czech Republic should be a continuation of the well working system of medical care for children with CHD. Specialized care for children with CHD is covered by one complex cardiac centre (Children's Heart Centre, University Hospital Motol, Prague) with a long-term stable outcome of 400–500 surgical interventions each year. This centre is directly connected to a nation-wide network of paediatric cardiologists and also closely cooperates with neonatology and paediatric units in the whole Czech Republic [10].

Follow-up observation of adult patients is to be similarly managed by a network of specialized centres for CHD in adults (CHD centres), complex cardiovascular centres (with a

**Table 1 – Diagnoses in adult patients with simple congenital heart disease.**

<b>Native disease</b>
Isolated congenital aortic valve disease
Isolated congenital mitral valve disease (e.g., except parachute valve, cleft leaflet)
Small atrial septal defect
Isolated small ventricular septal defect (no associated lesions)
Mild pulmonary stenosis
Small patent ductus arteriosus
<b>Repaired conditions</b>
Previously ligated or occluded ductus arteriosus
Repaired secundum or sinus venosus atrial septal defect without residua
Repaired ventricular septal defect without residua
From ACC/AHA 2008 Guidelines for the Management of Adults with Congenital Heart Disease [12].

specialized clinic for adults with CHD with 1–2 trained physicians), cardiology and general (internal) care units in regions, cardiologists in private practise and family practitioners. CHD centres should actively maintain this network, create working feedback with peripheral workers, offer easy access to consultations and cooperate on creating new arrangements for joint medical care.

The current European guidelines [1,11] do not strictly say which CHDs require medical care provided by CHD centres and which do not.

However it is considered ideal practise that every patient with CHD should be examined in a CHD centre upon reaching age of 18 years at least once. This should lead to a detailed medical report which should consist of a summary of the disease history including original diagnosis, past medical interventions, complications, patient's current clinical state, prognosis assessment and a plan for future follow-ups. In the more complex cases upon translation from paediatric cardiology to adult cardiology care it would be helpful to arrange a joint meeting with both a paediatric cardiologist and an adult cardiologist from a CHD centre being present.

To correctly determine which patients should be in the care of a CHD centre we could use a categorization based on complexity of the CHD [12]. All patients with CHDs of medium and high complexity should be at least partially monitored by a CHD centre. Only patients with simple CHD (Table 1) can be monitored by other cardiology institutions. In case of diagnostic or treatment uncertainty or complication any CHD patient could always be referred to a CHD centre.

### Statute of specialized CHD centres for adults

A supra-regional centre delivers super-specialized multi-discipline medical care for adult patients with CHD on the highest possible level and also provides teaching courses regarding CHDs. In order to do that such a centre needs to have adequate qualified staff, technical equipment and experience in the field.

### Staff requirements

Cardiologist experienced in CHD in adults	≥2
Imaging specialist (CMR, CT) experienced in CHD in adults	≥1
Congenital invasive cardiologist experienced in CHD in adults	≥2
Cardiac surgeon experienced in CHD in adults	≥2
Anaesthesiologist experienced in CHD in adults	≥2
Invasive electrophysiologist experienced in CHD in adults	≥1
Psychologist	≥1

Adequately experienced personnel are medical doctors with specialization certifications (in cardiology, cardiac surgery, radiology and imaging, anaesthesiology and intensive medicine) and at least a 2-year-long intensive training in a CHD centre. This arrangement ensures adequate erudition of personnel in management of CHDs and associated complications.

### Equipment requirements

ECG
Holter monitoring
Stress ECG
Ambulatory blood pressure monitoring
Event recorder
Cardiopulmonary exercise testing
Echocardiography (including transoesophageal echo, 3D echo)
CMR imaging
Cardiac computed tomography
Catheterization laboratory
Electrophysiology laboratory
Pacemaker/ICD implantation
Pacemaker/ICD after-care equipment
Cardiac surgery operating room

Additionally, a CHD centre needs to ensure a cooperation with an obstetric department in order to provide care for pregnant women with CHD. Other specializations that should be made easily accessible to adults with CHD are internal medicine (haematology, hepatology, nephrology, neurology), non-cardiac surgery, transplantology including long term ventricular assist devices and centres specializing in pulmonary hypertension. It is also necessary to maintain cooperation with geneticists, microbiologists, social workers, sports medicine doctors and lastly palliative care.

A smooth cooperation and transition between out-patients and in-patients care should be provided as well as a 24-h emergency service for urgent states in adult patients with CHD.

### Experience requirements

Expert committee for CHD in adults recommends that a minimum of 2000 patients should be in care of a CHD centre. Diagnoses of these patients should include medium and high complexity of CHD (CHDs with univentricular circulation, transposition of great arteries, Tetralogy of Fallot, Ebstein's anomaly, etc). This number reflects estimated low prevalence of complex CHD in population [13,3] and therefore only concentrating of these patients into specialized CHD centres will guarantee an adequate erudition of personnel regarding

medical care for medium and high complexity CHDs and even very rare CHDs. Such concentration of complex cases will create a solid knowledge base for solving a wide range of possible situations and complications in a heterogeneous population with CHD.

Moreover, a CHD centre needs to ensure an influx of new patients with CHD both from paediatric cardiologists and from regional cooperating cardiology practises. Using all their personnel, technical equipment and experience a CHD centre should be able to provide a range of all possible procedures required by adult patients with CHD.

In future we should aim to create a structured training and certification process for medical doctors working with adult patients with CHD.

According to information that was made available for the Expert committee for CHD in adults all the requirements for a CHD centre (including cardiac surgery for complex CHD in adults) are currently met by these institutions:

- *Centre for Adults with Congenital Heart Disease, Na Homolce Hospital Prague, Department of Cardiovascular Surgery [14]*
- *Centre for Adults with congenital Heart Disease, Motol University Hospital Prague (Department of Cardiovascular Surgery, Department of Cardiology, Children's Heart Centre, 2nd Faculty of Medicine, Charles University Prague)*  
both Prague Centres with connection to specialized care in the field of heart failure, long-term ventricular assist devices, transplantations and complex treatment of heart arrhythmias at the Institute for Clinical and Experimental Medicine.
- *Centre for Complex Care of Congenital Heart Disease in Adults – BRNO, University Hospital Brno and Centre for Cardiovascular Surgery and Transplantation Brno*

## Conflict of interest

None declared.

## Ethical statement

In agreement with the ethical codex of our institutions.

## Funding body

None declared.

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