Recommendations for standards of care

Grown-up congenital heart disease

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Introduction

Grown-up congenital heart disease (GUCH) is a rapidly growing field in adult cardiology with specific needs. Although most patients have undergone surgical repair or other interventional procedures in infancy or childhood, they are not cured and have residual haemodynamic and/or electrophysiological abnormalities. They are at increased risk of complications, thus requiring lifelong specialist medical care. The European Society of Cardiology (ESC) working group on GUCH has recently published guidelines for the management and practice recommendations for the standards and organisation of care for adults with congenital heart disease (CHD) [1, 2]. The Swiss Working Group for Adults and Teenagers with Congenital Heart disease (WATCH) has adapted these recommendations to the specific characteristics of the medical care system in Switzerland.

Goals

Given that GUCH cohorts are evolving and ageing, complex and changing treatment strategies are a particular challenge for many types of congenital heart defects in this patient population. A favourable impact on survival has recently been shown after the implementation of international guidelines and after referral of GUCH patients to specialised CHD centres [3]. In order to meet the needs of this special patient population and to establish adequate standards of care in Switzerland, recommendations for follow-up and institutional requirements need to be defined.

Key areas of attention in this position paper are therefore:

1. To define institutional requirements for regional and supraregional GUCH centres;
2. To define structured follow-up and establish minimal standards of care of GUCH patients.

Centres for grown-ups with congenital heart disease

GUCH centres are categorised as regional or supraregional centres. Regional centres work in close collaboration with a supraregional centre. All GUCH centres collaborate with paediatric cardiology services and have a structured transition process. Minimal infrastructural and staff requirements are outlined in table 1. GUCH cardiologists must have a nationally recognised certification in cardiology or paediatric cardiology with a minimal educational training of 24 months in GUCH. These 24 months should include at least 18 months in a supraregional GUCH centre and can be complemented by 6 months in adult cardiology for paediatric cardiology trainees and 6 months of paediatric cardiology for adult cardiology trainees [1].

Recommendations for structured follow-up and minimal standards of care for GUCH patients

Transition

- Every young adult with CHD should have a structured transition process beginning in teenage years. The paediatric cardiology team should provide adequate transition arrangements for patients and their families in collaboration with a GUCH cardiologist.
- At the time of transfer to the GUCH team, a comprehensive summary of the medical history (including operation reports from previous heart surgeries) must be provided in paper or electronic form.
- The time of transfer from paediatric to adult care is a vulnerable period and a clear plan for emergency situations around the time of transfer should be established.

Patient selection and follow-up recommendations

- Every adult with repaired or unrepaired congenital heart defect should be seen at least once in a GUCH centre, except those who have been discharged from

Table 1: Institutional and staff requirements for regional and supraregional GUCH centres (adapted from [1]).

	Regional GUCH centre	Supraregional GUCH centre
Minimal staff requirements:		
GUCH cardiologist(s)	1	2
Congenital cardiac surgeon(s)	–	2
Congenital invasive cardiologist(s)	–	1
Invasive electrophysiologist with GUCH expertise	–	1
Advanced cardiac imaging specialist with GUCH expertise	–	1
Minimal infrastructural requirements:		
GUCH outpatient clinic	+	+
24/7 emergency service	+	+
General adult medicine inpatient unit	+	+
Intensive care unit	+	+
Minimal requirements for collaboration with other specialities:		
Structured transition program with paediatric cardiology	+	+
Access to heart failure clinic	+	+
Heart transplant program	–	+
Multidisciplinary high risk pregnancy program	–	+
Access to dedicated clinical psychology / psychiatry	+	+
Access to genetic counselling	–	+
Access to palliative care	+	+
Access to social work supporting services	+	+

GUCH = grown-up congenital heart disease

follow-up by their paediatric cardiologist or those with a simple defect. Simple CHD is defined as a small atrial septal defect, ventricular septal defect or persistent ductus arteriosus, mild pulmonary valve disease or isolated aortic valve disease. If uncertainties in the management of patients with simple defects arise, all GUCH centres offer counselling about optimal care.

- Regular follow-up in GUCH centres is mandatory for all patients with non-simple CHD. We encourage a model of shared care between dedicated community-based cardiologists and GUCH centres, based on the patient's preference.
- All female GUCH patients contemplating pregnancy should be offered pre-pregnancy counselling at a GUCH centre. High-risk pregnancies should be managed by a dedicated multidisciplinary team.
- The intervals between clinical assessments and investigations must be individualised for each patient, based on lesion severity, importance of haemodynamic sequeale or residual lesions, estimated risk of complications or need for (re)-intervention.
- Follow-up recommendations for regular outpatient visits in adults with common non-simple CHD are as follows [2]:
  - atrioventricular septal defects (including ostium primum defects): at least every 3 years;
  - coarctation of the aorta: at least every 2 years;
  - Ebstein anomaly: at least every 2 years;
  - repaired tetralogy of Fallot: at least every 2 years;
  - arterial switch for transposition of the great arteries: at least every 2 years;
  - subpulmonary ventricle to pulmonary artery conduits (e.g. after Rastelli repair): at least every year;
  - subaortic right ventricles (e.g. after atrial switch procedure for transposition of the great arteries): at least every year;
  - Fontan palliation: at least every year;
  - cyanotic heart disease including Eisenmenger physiology: at least every year.

It needs to be emphasised that these are recommendations for minimal follow-up in low-risk patients within each diagnostic group. For patients at higher risk or during pregnancy, closer follow-up is required.

Cardiac and noncardiac intervention in GUCH patients

- All GUCH patients with non-simple CHD requiring structural cardiac interventions (e.g., percutaneous or surgical valve implantation) should be treated in a supraregional GUCH centre where the expertise of a congenital cardiac surgeon or invasive cardiologist is available. Electrophysiological procedures should be performed by a dedicated electrophysiologist with experience in CHD in close collaboration with the patient's GUCH cardiologist.
- Noncardiac interventions for cyanotic patients and patients with single ventricle physiology must be performed in GUCH centres. For all patients with non-simple lesions requiring noncardiac interventions we recommend pre-interventional consultation with the patient's GUCH cardiologist.

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A full list of references is available in the online version of this article.

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