

ESC 2015 – Highlights in Grown-up Congenital Heart Disease and Congenital Cardiology
An overview provided by Nucleus of the Working Group on Grown-up Congenital Heart Disease for
the members of the Working Group.

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on behalf of the Nucleus of the Working Group on GUCH.*

The ESC Congress 2015 in London, GB attracted 32,778 participants from around the globe. It hosted 22 sessions related to congenital heart disease and closely related fields (such as pregnancy and heart disease). The program included more than 150 contributions (both oral presentations and posters) in the field. Of these more than 100 presented novel, original data.

The aim of this newsletter is to summarize important contributions in the field of grown-up congenital heart disease/congenital heart disease presented at the meeting. It highlights what the authors perceive to be the highlights of the congress. While we aim to provide a comprehensive and balanced overview of the area, the selection and presentation is by necessity a subjective one. Therefore, many worthwhile contributions could not be included here. For a full overview of the scientific program in the field please refer to the ESC website (www.escardio.org). In addition, ESC365 (a free service for ESC members) provides the presentation slides and recordings of many sessions.

ESC 2015 - Original Contributions Congenital Heart Disease

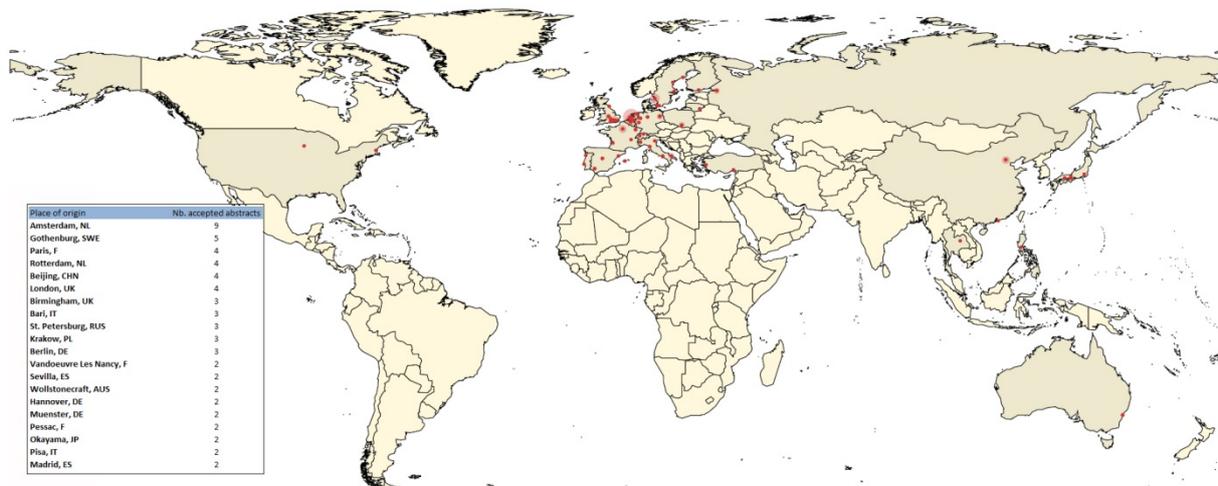


Figure: Origin of original contributions (scientific abstracts/posters) by country and city. All cities with more than 1 accepted contribution are listed. The size of the red dot corresponds to the number of contributions. Countries with active participation are in light orange.

General Trends and Overview

As in previous years, many (un-)natural history studies and retrospective studies were presented. It appears that the trend to ever larger datasets continues. In addition to contributions from established large single centres and well known national registries (such as the Dutch CONCOR and the German National CHD Register), population-level studies based on administrative datasets are increasingly represented and provide an alternative view on the area. These large datasets, often linking various data sources, provide a direct real-world insight into prevalence of disease and outcome in the community. This greater scope comes at the price of reduced depth of information, with clinical details often missing. It seems, however, that *Big Data* has reached GUCH and is here to stay. As this type of data is difficult to handle and analyse it will continue to pose challenges to academics working in the field and acquiring new skills may be required for many of us.

Pregnancy continues to be of major interest as half of our adult patients are female and many plan to have a family. Appropriate counselling and management of pregnancy continues to be a challenge: Beyond, risk stratification novel insights into the pathophysiology of pregnancy in congenital heart disease patients have been presented.

Improved imaging modalities and the use of (novel) biomarkers has also attracted interest, which could be included as part of better risk stratification tools in the future.

Also, as a consequence of improved outcome, the focus is increasingly shifting from mortality to morbidity, quality of life and functional outcomes while pulmonary hypertension remains in the focus of attention as in previous years.

Natural History Studies

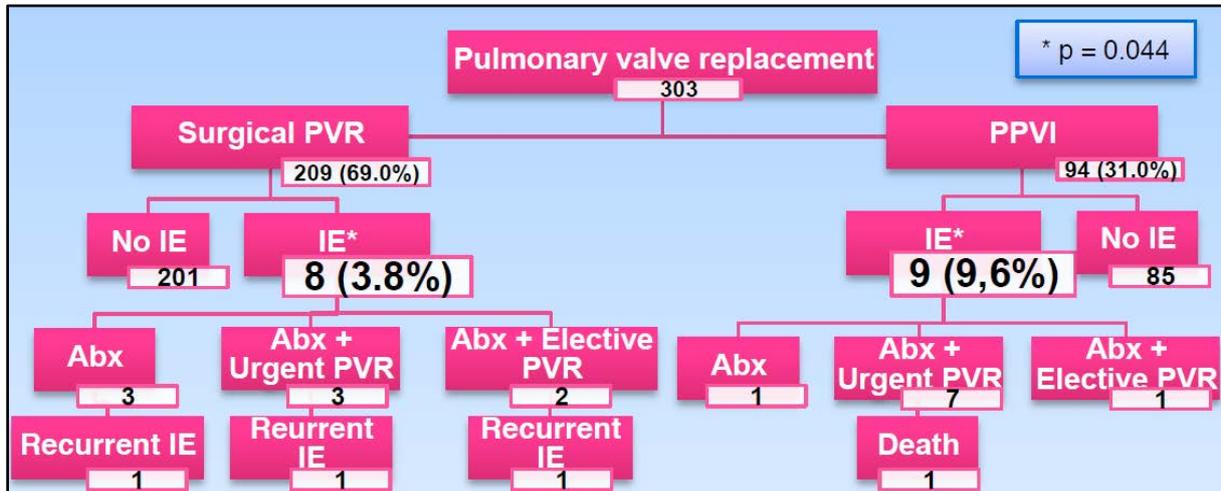
[Dr Dellborg \(P2654; Gothenburg, SWE\)](#) presented data on survival trends in children born with congenital heart disease in Sweden between 1970 and 1993. This study is based on Swedish patient-databases linked to cause of death registers. The authors included data on 21,564 patients with congenital heart disease. After a mean follow-up of 13 years 93.1% of patients were still alive and improvements in outcome over time were evident. **Source:** European Heart Journal (2015) 36 (Abstract Supplement), 457

[Drs Rosengren and Mandalenakis \(P2657 and P438; Gothenburg, SWE\)](#) also presented data on the risk of stroke in patients with congenital heart disease based on the Swedish registries mentioned above. They report an increased risk of, both, ischaemic and haemorrhagic strokes in this population, highlighting the increasing importance of non-cardiac disease in this growing and aging cohort. **Source:** European Heart Journal (2015) 36 (Abstract Supplement).

[Dr Cuypers \(P 2656; Rotterdam, NL\)](#) presented data on mortality and morbidity in a longitudinal followed cohort of patients who underwent surgical correction for pulmonary stenosis at young age. She found that survival up to 40 years after successful repair of PS was nearly as good as survival in the general population. Subjective health status was good and there was a low incidence of arrhythmias. Re-interventions, however, were necessary in one quarter of the patients. **Source:** European Heart Journal (2015) 36 (Abstract Supplement), 457

Endocarditis

[Dr Cesna \(P2141; Vilnius, LT\)](#) investigated the important topic of endocarditis related to percutaneous pulmonary valve replacement (PVR). Based on 303 cases of pulmonary valve replacement (69% surgical, 31% interventional) the authors report a significantly higher rate of infective endocarditis in interventional PVR (9.6%) compared to patients post-surgical PVR (3.8%). This finding deserves further study and – as suggested by the authors – careful patient assessment (and possibly appropriate counselling) before interventional procedures.

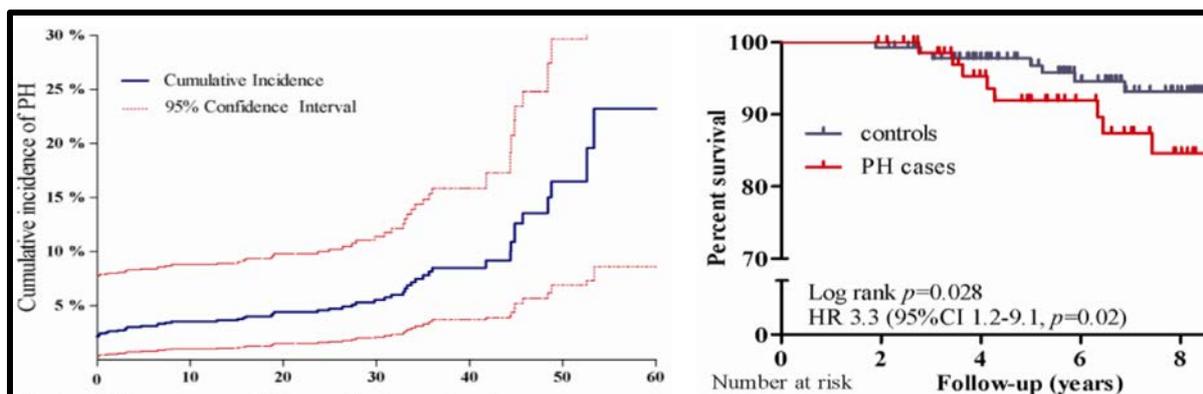


Source: European Heart Journal (2015) 36 (Abstract Supplement), 357

[Dr Kuijpers \(A2934; Amsterdam, NL\)](#) used the Dutch CONCOR database to assess the incidence and predictors of infective endocarditis in GUCH patients. Based on a patient population of 15,284 patients, the authors identified a history of endocarditis in 363 patients (2%). Predictors of endocarditis were multiple defects, the presence of prosthetic material (especially valves) and male gender. Interestingly a previous history of endocarditis did not emerge as a risk factor on multivariate analysis. This study suggests the possibility that valvar prosthetic material should be considered as a main risk factor for endocarditis. Source: European Heart Journal (2015) 36 (Abstract Supplement), 515

Pulmonary Hypertension in Congenital Heart Disease

[Dr Van Riel \(P437; Amsterdam, NL\)](#) challenged the dogma of curative shunt closure at “young” age. Based on the well-established Dutch CONCOR Registry she and her co-workers assessed a random sample of 1,877 patients born with systemic-to-pulmonary shunt lesions. These patients were carefully reviewed. The authors report that 1,103 patients underwent shunt repair, but despite surgery 72 patients still developed PH during long-term follow-up. Alarmingly, cumulative incidence of PH at the age of 50 years exceeded 15% in this population (left panel). In addition, PH patients had significantly worse survival prospects compared to controls without the condition (right panel).

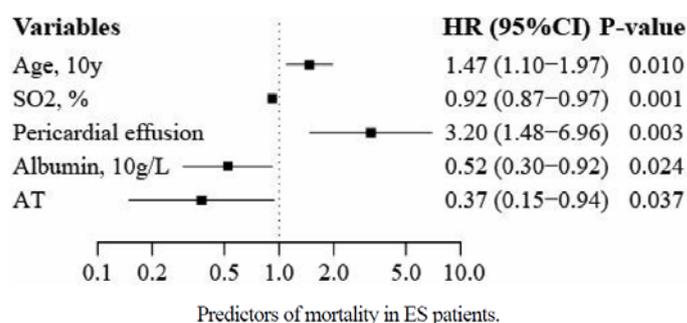


Source: Abstract P437 (EHJ 2015, 36 Abstract Suppl. 36-37) and J Am Coll Cardiol. 2015;66(9):1084-6.

As a consequence, this study suggests that despite repair at “early” age GUCH patients with previous left-to-right shunt lesions (including ASDs) require lifelong follow up and proactive treatment of PH may be warranted. **Source:** European Heart Journal (2015) 36 (Abstract Supplement), 357

[Dr Blok \(P2658; Amsterdam, NL\)](#) presented new data on the use of Macitentan (a novel endothelin receptor antagonist, ERA) in patients with pulmonary arterial hypertension (PAH) associated with congenital heart disease or Eisenmenger syndrome. This was a prospective observational study including a protocolized approach with regular 6-minute walk test assessment. The authors report that most patients (74% of whom had Eisenmenger syndrome) improved after being switched from Bosentan (another ERA) and this was accompanied by reductions in NT-pro BNP levels. It is concluded that a switch from Bosentan to Macitentan appears to be safe and to improve clinical outcome (albeit 6-minute walk test distance remained stable). **Source:** Abstract P2658 (EHJ 2015, 36 Abstract Suppl. 458).

[Dr Kempny \(P6417; London, GB\)](#) presented new data on risk stratification in patients with Eisenmenger syndrome. Based on analysis of 334 adult Eisenmenger (ES) patients, the authors were able to identify a number of simple clinical parameters that correlated well with prognosis on multivariate analysis. These variables include patient age, resting oxygen saturation, albumin levels, the presence of pericardial effusion on echo and the fact that the patient is treated with advanced therapies (see Figure below):



Source: European Heart Journal (2015) 36 (Abstract Supplement), 1138

[Dr Blok \(P2661; Amsterdam, NL\)](#) presented the results of a prospective observational study aiming to identify prognostic markers in pulmonary hypertension associated with congenital heart disease. The authors included not only well established clinical parameters and 6-minute walk test but also novel biochemical markers such as NT-pro BNP and cystatin C. The authors were able to demonstrate that NT-pro BNP and cystatin C (a marker of renal function) outperformed conventional clinical parameters and surprisingly also 6-minute walk test distance regarding their prognostic value. This topic certainly deserves further study as risk stratification remains an unsolved issue in this challenging population. **Source:** European Heart Journal (2015) 36 (Abstract Supplement), 458-459

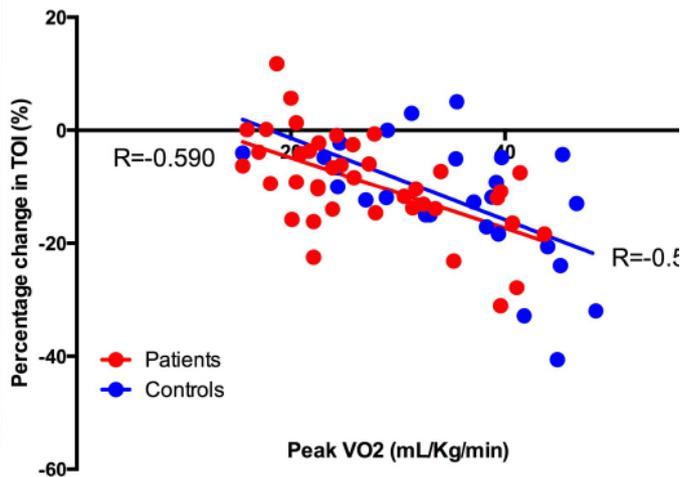
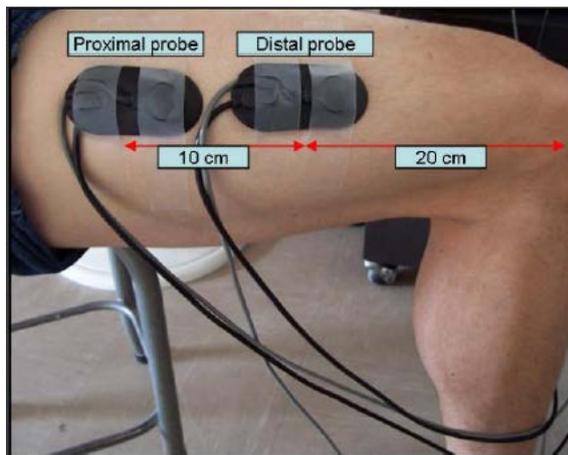
Educational Lectures:

Advanced management of pulmonary hypertension in congenital heart disease was the topic of a session chaired by Prof. Gomez Sanchez (Madrid, ES) and Prof. Budts (Leuven, BE). The slides and recordings of the lectures are available on ESC365. These include the topics of: *Natural history and progression of pulmonary hypertension in congenital heart disease* (Diller, DE); *Predicting deterioration and need for escalation of treatment* (D'Alto, IT); *Experience with intravenous, subcutaneous and inhalative therapies* (R Alonso Gonzalez, GB) and *Pregnancy and contraception in pulmonary hypertension and congenital heart disease* (Roos-Hesselink, JW).

Functional assessment and exercise capacity

[Dr Sandberg \(P2659; Umea, SWE\)](#) showed data from a prospective study examining the effect of home-based exercise training on maximal exercise capacity in adults with moderate complexity/complex congenital heart disease. 17 patients were randomized 2:1 to 12 weeks of home based prescribed level ergometer cycle training or usual activity. The authors report that peak workload and exercise time improved as a result of the training program. **Source:** European Heart Journal (2015) 36 (Abstract Supplement), 458

[Dr Frigiola \(P2139; London, GB\)](#) presented an important insight into the relative contribution of central and peripheral factors in determining exercise capacity in tetralogy of Fallot (ToF) patients. It is well appreciated that these patients have reduced exercise capacity. Frigiola and co-workers used near infrared spectroscopy (NIRS) measurements at the vastus lat. muscle to assess the importance of peripheral/skeletal muscular factors for exercise capacity (left panel). In addition, patients also had cardiac MRI data available. Interestingly, a strong correlation between peripheral muscular oxygen extraction on NIRS and peak oxygen consumption was found (right panel). In contrast, cardiac MRI parameters of ventricular function (at rest) were not correlated with exercise capacity. This important study highlights that patients with simple or moderate complexity lesions may benefit from exercise training/cardiac rehabilitation more than commonly appreciated and that resting haemodynamic parameters are a poor predictor of exercise capacity in this population (and generally in CV disease).



Source: European Heart Journal (2015) 36 (Abstract Supplement), 357

Pregnancy

[Dr Banerjee \(P712; Birmingham, GB\)](#) presented an interesting study assessing risk of aortic dissection during pregnancy in England based on national databases. Their study illustrates the great potential but also highlights the challenges of using large and diverse administrative databases to this end. **Source:** European Heart Journal (2015) 36 (Abstract Supplement), 109.

[Dr Orwat \(A143; Muenster, DE\)](#) presented the outcome of women with moderate or severe aortic stenosis undergoing pregnancy. Based on the unique ROPAC Registry, he and colleagues assessed the outcome of 99 pregnancies in 96 women (34 with severe aortic stenosis). Reassuringly maternal mortality in this study was 0% but symptomatic and esp. severe aortic stenosis carried a substantial risk for heart failure, hospitalisation for cardiac reasons and preterm birth/low birth weight in up to 1/3 of patients. **Source:** European Heart Journal (2015) 36 (Abstract Supplement), 5.

[Dr van Hagen \(A142; Rotterdam, NL\)](#) presented a study on risk assessment in pregnant women with structural heart disease. Also based on the ROPAC Registry, she tested the mWHO risk stratification model in both developed and developing countries. It became clear that complications occurred more often in developing countries. The modified WHO risk classification was confirmed as a useful tool for predicting cardiac events during pregnancy. Originating from a developing country and having atrial fibrillation were found to be independently associated with a higher risk. **Source:** European Heart Journal (2015) 36 (Abstract Supplement), 4-5

[Dr Kampman \(P713; Groningen, NL\)](#) presented an interesting prospective and mechanistic study assessing uteroplacental circulation in pregnant women with tetralogy of Fallot (ToF). The authors investigated 55 ToF women and show that ToF patients had more often abnormal uteroplacental Doppler flow (UDF). Furthermore, abnormal UDF was linked to maternal right ventricular dysfunction and was associated with offspring complications (low birth weight). **Source:** European Heart Journal (2015) 36 (Abstract Supplement), 109-110.

Fontan circulation

[Dr Schwerzmann \(A2929; Bern, CH\)](#) presented the results of an interesting prospective (FALCON) study investigating the impact of short-term exposure to low barometric pressure (equivalent to an altitude of 3400 metres) on exercise capacity and haemodynamics in Fontan patients. This topic is of major interest as hypoxia may lead to pulmonary vasoconstriction and theoretically subsequent Fontan failure. Using research facilities at low level and on the Jungfrauoch mountain in Switzerland (3400 metres above sea level), Schwerzmann and co-workers performed cardiopulmonary exercise tests and measured pulmonary blood flow (PBF) non-invasively in 17 adult Fontan patients and 15 matched healthy controls. They found that – despite theoretical concerns – Fontan patients showed good adaptation of PBF to high altitude with no negative impact on PBF or exercise capacity when compared to healthy controls.

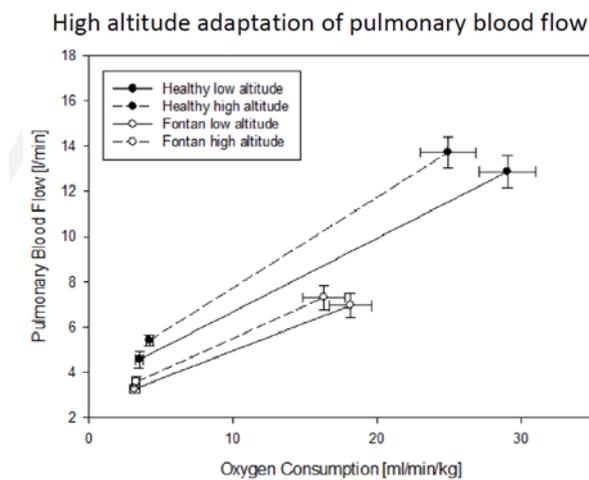


Figure: Compared to controls, Fontan patients showed comparable change in PBF and a relatively even lower reduction in oxygen consumption.

Source: European Heart Journal (2015) 36 (Abstract Supplement), 514

Tetralogy of Fallot (ToF)

[Dr Bokma \(P2137; Amsterdam, NL\)](#) and colleagues investigated the prognostic value of a fragmented QRS complex in patients with ToF. Based on a Dutch nationwide registry they included 794 patients and classified QRS fragmentation as absent, moderate or severe. During a follow-up time of 10.4 years (median) 46 patients died and 28 had a sustained ventricular tachycardia. Fragmented QRS complexes (esp. severely fragmented QRS) emerged as an independent predictor of outcome and was prognostically superior to QRS duration in this study.

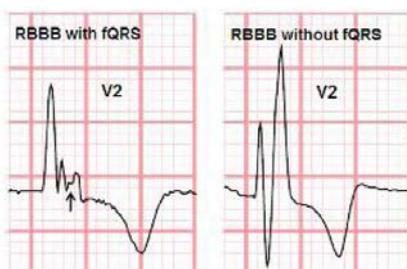


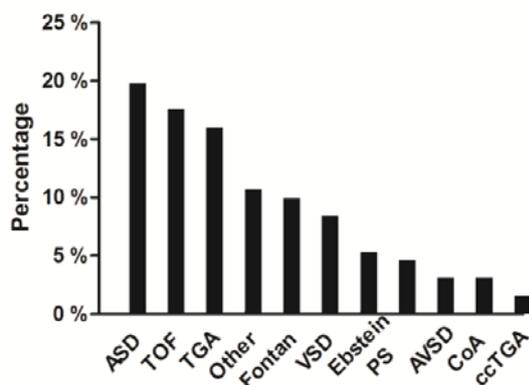
Figure: Example of fragmented QRS complex.

Source: European Heart Journal (2015) 36 (Abstract Supplement), 356

[Dr Vormbrock \(P2138; Münster, DE\)](#) investigated whether an elevated LVEDP, a recognized prognostic marker in patients with ToF reflects right ventricular pathology or represents a marker of intrinsic LV disease. Including 40 ToF who underwent cardiac catheterization and cardiac MRI, the authors report that LVEDP did not correlate with RV volumes or function. Rather LVEDP was weakly correlated to systemic arterial pressure, suggesting that LV filling pressures represent a marker of intrinsic LV dysfunction in patients with ToF. **Source:** European Heart Journal (2015) 36 (Abstract Supplement), 356-357

Anticoagulation - NOACs

[Dr Yang \(A2927; Amsterdam, NL\)](#) gave an update on the ongoing prospective multicentre registry of NOAC use in adult congenital heart disease. She reported that so far 133 patients have been included. The distribution of defects is presented in the left panel. Preliminary results suggest that events of thromboembolism and major bleeding with NOACs may be at least comparable to those under vitamin K antagonists (VKA), although further analysis is required (right panel).



ACHD with AA	Annual events	
	VKA	NOAC
Thromboembolism	1.4%	0%
Major bleeding	4.4%	1.1%

Source: European Heart Journal (2015) 36 (Abstract Supplement), 513

Acquired Cardiovascular Disease in GUCH

[Dr Koerten \(P724; Berlin, DE\)](#) screened the National Registry of Congenital Heart Defects for patients with established cardiovascular risk factors. Among 540 patients with congenital heart disease he and coauthors identified a relatively large proportion of patients with known cardiovascular risk factors (e.g. arterial hypertension in 20.9% of pts., diabetes in 10.6% and hyperlipidaemia in 15.0%). Alarming, despite being frequent, these risk factors were insufficiently treated in many patients (e.g. only 2.1% of patients had statins and only 66.7% of pts. with arterial hypertension were appropriately treated). **Source:** European Heart Journal (2015) 36 (Abstract Supplement), 356357

Additional educational lectures of potential interest

The following interesting lectures are available on ESC365:

Surgical ablation of ventricular arrhythmias in congenital heart disease.
Session: Challenges in treating arrhythmias in adults with congenital heart disease
Presenter: A Giamberti (San Donato Milanese, IT)

The dilated ascending aorta after repair of complex congenital heart disease.
Session: Surgical challenges in the adult with congenital heart disease
Presenter: H Baumgartner (Muenster, DE)

Treatment of atrial arrhythmias in congenital heart disease.
Session: Challenges in treating arrhythmias in adults with congenital heart disease
Presenter: C Berul (Washington, US)