both arms. Although the pulses were equal over both upper extremities significant radial-femoral delay was noted. The bilateral femoral and popliteal pulses were found to be nonpalpable; 2/6 midsystolic murmur was heard on the left scapular region in the back. High voltage were encountered on the resting ECG. Transthoracic echocardiography showed concentric left ventricular hypertrophy accompanied in spite of the fact that there was found no significant pressure gradient distal to aortic isthmus in doppler analysis.

At subsequently performed cardiac catheterisation via right brachial and femoral arteries, a complete aortic arch interruption distal to the subclavian artery take-off was imaged using a simultaneous approach from the right brachial artery and femoral artery (Figure 1A). The peak systolic pressure above the interrupted segment was 190 mmHg, and the pressure below it was 100 mmHg. Then, the occlusion was perforated retrogradely from descendin aorta by way of the right femoral artery using coronary chronic total occlusion guidewire (Hi-torque Pilot 200 190 cm, Abbott Vascular, USA) that was snared with a Amplatz Goose neck snare (eV3 company, USA) from the right brachial artery. Over an brachio-femoral arterial loop performed using an a 0.014" coronary guidewire, the stenosis was consecutively dilated with coronary (1.5.0X20 mm, 3.5X20 mm; brio balloon, CID Company, Italy) and peripheral angioplasty balloons (6.0X20 mmViatrac 14 Plus; Abbott Vascular, USA). Then, a 12 F delivery sheath crossed the dilated segment of the aorta; subsequently, a 28 mm long covered Cheatham Platinum (CP) stent (NuMED Inc, New York, USA) was successfully relieved by progressively dilated to 24 mm (Figure 1B). Post-procedure peak pressure gradient was 5 mm Hg. After 3 months, control echocardiography showed 30 mmHg maximal gradient. Control angiography demontrated a maximal gradient of 32 mmHg. We performed second aortic balloon angioplasty into the stent; subsequently, maksimal gradient decreased to 5 mmHg (Figure 1C).

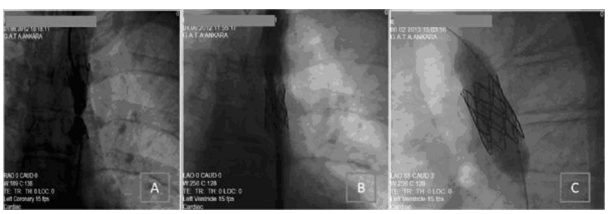

Figure 1. Aortic angiography in postero-anterior view from brachial artery and femoral artery imaging a complete interruption of the aortic isthmus (Figure 1A). Aortogram in lateral view after stent implantation, showing complete relief of the aortic interruption (Figure 1B). The patient underwent second balloon angioplasty (Figure 1C).

PP-369

Modern Computed Tomography in Congenital Heart Disease.
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Objectives: Modern imaging technologies are essential in the pre-, peri- and postoperative management of patients with congenital heart disease. Besides echocardiography, computer tomography (CT) and magnet resonance imaging (MRI) are gaining importance. The aim of the present study was to show indications, options and limitations of multislice-CT (MSCT) in congenital heart disease.

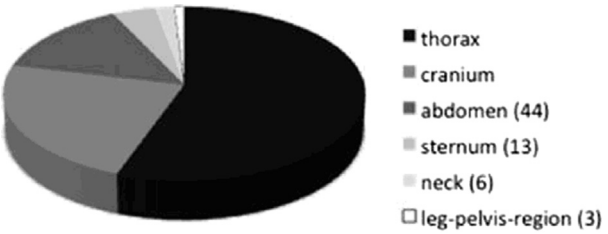

Figure 1. Regions of interest for MSCT diagnostic.

Methods: Data of all consecutive patients who underwent an MSCT was retrospectively collected over 32 months. Timing of MSCT (pre-, peri- or postprocedural) was also reflected. Indications for MSCT diagnostic were divided in: thoracic, cerebral, abdominal, skeletal, neck sternum and leg-pelvis pathologies.

Results: A total of 250 CT scans were performed in 195 patients (84 women; median age 22 years). Seventy-five patients had complex or rare congenital heart disease. According to the evaluated region, diagnosis was divided into: intrathoracic structures (n=172), cerebral (n=72), abdomen (n=44), sternum (n=13), neck (n=6) or leg-pelvis region (n=3). Questions answered were exclusion/confirmation of pulmonary embolism (n=29), aortic aneurysm/dissection (n=22), clarification of cerebral symptoms (n=40), sternal disorders (n=12), inflammation (n=27), choanal atresia (n=3), haemoptysis (n=4), vascular anomalies (n=3) and others (n=10). In 38 patients more than one CT was necessary. A diagnosis was achieved in 94% of cases.

Conclusions: MSCT diagnostic is a helpful tool in the perioperative management and long-term follow-up of patients with CHD. Extracardiac findings are frequent. A strong cooperation between congenital cardiologists and experienced radiologists is mandatory.

PP-370

Coagulation Disorders in Marfan Syndrome.
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Objectives: Aortic complications are the main cause of death in Marfan syndrome but coagulation disorders may play a role in progression and

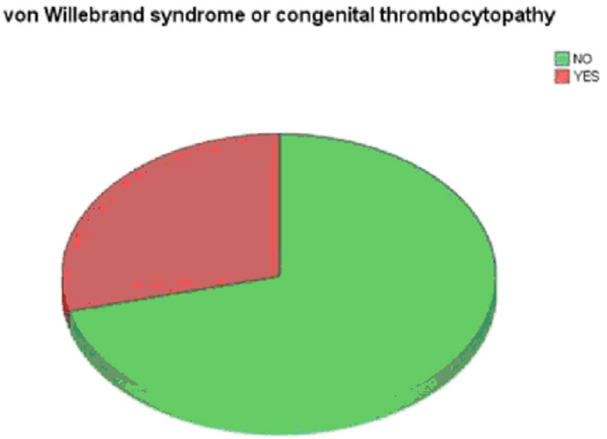

Figure 1. Suspected von Willebrand syndrome or other congenital thrombocytopathies in Marfan patients.

POSTER ABSTRACTS

outcome of aortic disease in these patients. The aim of this study was to investigate coagulation disorders in a prospective Marfan cohort.

Methods: Blood samples were collected in consecutive patients with verified Marfan syndrome who attended our outpatient clinic from December 2012 to October 2013. We analysed whole-blood platelet aggregation, aPTT, fibrinogen, D-dimers, Prothrombinfragment 1+2 (F1+2) and calibrated automated thrombogram (CAT).

Results: 38 consecutive patients with proven Marfan syndrome (50% women, mean age 37.4 ± 11.5 years) were enrolled. Pathologic D-dimer levels were statistically significant in 5 patients, all of them with chronic dissection of the thoracic or abdominal aorta (p= 0.001, respectively). Fibrinogen was increased in patients with aortic regurgitation (p= 0.03) and there was a positive, significant correlation between age and fibrinogen (R= 0.43, p= 0.006). In vitro bleeding times (PFA-ADP-Collagen and PFA-Epinephrin-Collagen induced platelet function) were prolonged in 11 patients (28.9%, p= 0.009), which is compatible with von Willebrand syndrome or congenital thrombocytopathy. In addition, F1+2 was elevated in 9 patients, whereas CAT results were decreased mostly.

Conclusions: Coagulation plays a major role in Marfan syndrome. Both procoagulation and bleeding disorders participate in the disease. Platelet function abnormalities, compatible with von Willebrand syndrome or congenital thrombocytopathies, have been identified in 29% of patients. The results of different thrombin generation methods deserve further evaluation.

As surgical procedures are often necessary in patients with marfan syndrome, the assessment of the coagulation status should be given greater attention.

PP-371

Noncardiac Comorbidities of Congenital Heart Disease in Adults.

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Background: Congenital heart anomalies (CHD) may be complicated by noncardiac comorbidities (NCCO), which may modify or aggravate the natural, the postoperative or the postinterventional course of the disorder.

The aim of this prospective study was to evaluate the proportion of significant noncardiac illnesses in adults with CHD.

Methods: In one tertiary care center for CHD 830 consecutive adults, representing all types and severity grades of acyanotic and cyanotic CHD (age: range 18 − 79 ys; 56 % female), were identified during a 1-year-period and evaluated for significant acquired comorbidities.

Results: Associated problems were diagnosed in most patients (n = 830; 95 %). NCCO mainly stemmed from the field of respiratory medicine, gastroenterology/hepatology, nephrology, endocrinology, gynecology/obstetrics, neurology/psychiatry, hematology, otorhinolaryngology, orthopaedics, and dermatology. More than 10 % had a genetic or syndromic disorder. At least 74 % of all patients were on a chronic cardiac and/or non-cardiac medication. Laboratory findings of renal or hepatic disorders in 625 patients gave evidence of renal involvement in more than 6 % and hepatic involvement in 10 %.

Conclusion: A considerable proportion of adults with CHD has significant noncardiac comorbidities, which may profoundly influence the natural course of the disease, the prognosis, the treatment strategies and the upcoming costs. Moreover, dysfunction of organ systems such as liver, kidney, coagulation system, or thyroid gland, may disturb drug action. In order to understand all organ-specific aspects and to improve thus the long-term care for these patients close collaboration between congenital cardiologists and other subspecialties is crucial.

PP-372

Characteristics of Adults with Congenital Heart Disease and Pulmonary Arterial Hypertension in the COMPERA-CHD Registry.

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Background: About 90% of the patients with congenital heart defects (CHD) reach adulthood today. In app. 10% of them pulmonary arterial hypertension (PAH) exists and adversely affects exercise capacity and prognosis. Main aim of the study is to document real-life data of adults with PAH.

Methods: In the COMPERA registry (Prospective Registry of Newly Initiated Therapies for Pulmonary Hypertension, ClinTrials.gov Identifier NCT01347216) adults with pulmonary hypertension are documented since 2007, if they are treated with disease-specific drugs (e.g. endothelin receptor antagonists, phosphodiesterase-5-inhibitors, prostacyclins, newer specific drugs). Further included are untreated patients with Eisenmenger syndrome, as well as patients with pulmonary vascular dysfunction (modified Fontan operation).

The registry documents medication (mono- or combination-therapy), continuity of treatment, change in therapy, adverse events, treatment effects (in 6-minute walk test, functional class).

Results: As of September 2013, a total of 4.515 PAH/PH patients were documented in 30 centres in 7 countries. 378 had PAH-CHD (8 %; age 46.5 ± 17.0; 36 % males; NYHA functional-class (FC) I/II in 31.2 %, FC III in 64.5 %, FC IV in 4.2 %; mean 6-minute walking distance 364 ± 119 meters; quality of life 50.4 ± 23.3 points. Detailed information from a special CHD case report form in 180 patients (168 Eisenmenger, 12 Fontan) is presented in the following: The majority of these patients were treated with PAH specific drugs, usually as monotherapy In the Eisenmenger-group (n = 168) 71 % were on monotherapy, 17 % on combination therapy, 12 % without PAH-specific drugs. In the Fontan-group (n = 12) all were on monotherapy. ERA were given more often than PDE-5 inhibitors, prostacyclines only exceptionally.

In the entire Compera-CHD-cohort (n=322) who received PAH-specific therapy, 34 patients died during a follow-up of 5 years. The mortality rate of CHD-patients is with 10.6 % significantly lower - compared to 19.4 % (n = 221) in idiopathic PAH (IPAH) (p=0.0002, simpler Chi-Quadrat Test). The 5-year (Kaplan Meier) survival estimate during follow-up was 84 % in ES, and 62 % in IPAH (p=0.000001).

Conclusion: Current COMPERA data indicate that CHD patients