LVOT gradient of 200 mmHg). These echocardiographic and cutaneous manifestations together with deafness were present prior to ECG alterations and marked clinical symptoms. At the age of 14 years of operation treatment of HCM had to be performed due to clinical symptoms including syncope and rapidly progressive cardiac alterations.

Conclusion: PCL is a cardiovascular syndrome which rarely has been reported. However, being aware of these characteristic cutaneous alterations, 3 pts with PCL were newly diagnosed in our clinic in the course of 1 year. In pts with multiple linear telangiectases a thorough cardiac assessment has to be performed. If no cardiac abnormalities are detected by TTE, repeated cardiac reassessment (TTE) should be performed as HCM may develop later and may be rapidly progressive. Cardiologists should be aware of characteristic skin alterations leading to the early diagnosis of cardiomyopathy (neurofibromatosis, lentigines and angiofibromas in Fabry disease) for prognostic and therapeutic reasons.

Echocardiographic aspects in beta thalassemia major

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Aims: This is a prospective study intending to determine the echographic parameters (transsthoracic examination) at the patients with beta thalassemia major and multiple blood transfusions.

Background: Multiple transfusions, clinically beneficial for the patients with beta thalassemia major cause iron overload and myocardial function involvement (heart failure is often present before 20 years). Cardiac damage leads to a type of cardiomyopathy, with both systolic and diastolic dysfunction, frequently associated with arrhythmias.

Methods: Patients eligible for this study are those referred to our center after blood transfusions and are investigated with transsthoracic echocardiography. The measurements made are: LVID diameter, LA diameter, LV wall thickness, LV systolic and diastolic function (the last, with pulsed Doppler used to measure LV inflow through the mitral valve). 16 patients are included (11 male; age 6-23 years, median 12.7 years) in 2003-2005. All of them presented moderate anemia (Hb>7g/dl) and markedly elevated values for serum ferritin, with the use of the chelating agent 6 of them had splenectomy and 2 arrhythmias (supraventricular and ventricular tachycardia).

Results: It was found LVWV enlargement in all patients and for 11 pts. at LA too. Most patients had increased LV wall thickness (10-15 mm mean). LV ejection fraction was preserved in all cases. E/A ratio (normal between 1.8 and 2.5 at children) was >2.5 at 3 pts. >2 at 5 pts (median 1.76). For 3 patients rehospitalised, we found at 1, respectively 3 and 10 months, that the pattern is changed: from delayed relaxation to restrictive-2 and from restrictive to pseudo-normalization.

Conclusions: In advanced stages of beta thalassemia major, the pattern of cardiomyopathy is mixed, restrictive and dilated. Cardiac involvement in this disease leads to LV diastolic dysfunction, the systolic function is preserved till late in evolution. Frequently, the ventricular wall thickness is increased, myocardial damage being due to direct tissue toxicity of the free iron moiety and tissue infiltration. The LVM reduction in otherwise asymptomatic patients following treatment with iron chelation therapy could be best seen in 2 and 4 chamber views. In one case, in addition to classical LV hypertrophy, was present a thick myocardial bundle along interventricular septum. In 4 pts (3 younger than 40 years) LV systolic function was normal (EF >50%), in one EF was 50%. In 2 pts there was LV systolic dysfunction with EF between 25% and 45%. All pts older than 40 years had abnormal LV systolic function. In 2 pts with impairment of LV and severe mitral regurgitation replacement of the valve was necessary, in other 2 mitral insufficiency was mild. 3 young pts had mitral valve prolapse. One pt had congenital fistula between left coronary artery and pulmonary artery. There were no thromboembolic events.

Conclusions: In adult patients with myocardial noncompaction systolic LV dysfunction is common, predominantly in older persons; sometimes with severe mitral regurgitation, in this cardiomyopathy ventricular arrhythmias are a common cause of sudden cardiac death.

Stress-induced apical ballooning syndrome can be diagnosed early based on classic echocardiographic findings

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Background: Stress induced apical ballooning has been described as a reversible condition involving the apical left ventricular wall, sparing the base, and causing a ballooning appearance of the left ventricle during systole despite normal coronary arteries. We observed early diagnosis of this syndrome based on classic echocardiographic findings that involves many coronary territories with apical ballooning appearance.

Method: We are presenting four cases of apical ballooning syndrome that was seen at our institution. We evaluate echocardiographic characteristic of this syndrome that could lead to early diagnosis before cardiac catheterization.

Results: All Echocardiograms showed similar anatomical apical ballooning of the left ventricular apex. The diagnosis of apical ballooning syndrome was suspected based on echocardiography in conjunction with clinical data before cardiac catheterization was performed. All cases revealed mid to apical involvement of the walls of the left ventricular apical territories with apical base and ballooning appearance that could be best seen in 2 and 4 chamber views. In one case, in addition to classic left ventricular apical ballooning, marked right ventricular apical akinesia was present on the initial echocardiographic examination. Subsequent angiograms in all these patients showed classic apical ballooning appearance during ventriculographic examination with normal coronary arteries.

Conclusion: Apical ballooning syndrome can be suspected based on classic echocardiographic findings and clinical data prior to angiography that could lead to early treatment and intervention of this syndrome.

Functional mitral regurgitation is associated with the degree of aortic stiffness

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Functional mitral regurgitation (FMR) is strictly related to ventricular afterload.