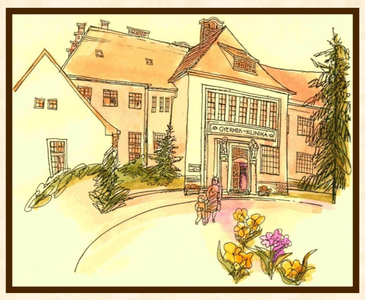

Pediatric myocarditis: new aspects from a case series

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Background and Aims:

Our knowledge regarding the clinical characteristics of myocarditis presenting in childhood is patchy and inconclusive. Prolonged follow up created an opportunity to gather additional important knowledge regarding the nature of this rare condition. The clinical characteristics of paediatric myocarditis treated in our institute were analyzed in order to generate new knowledge about the course of the illness and possible comorbidities.

Methods:

A retrospective data collection was performed to identify patients with myocarditis treated in our institute between the 1st of January 1996 and 31st of December 2010. The median follow up time was 12.71 years (range: 3.57-15.2 years).

Myocarditis was defined as severe cardiac dysfunction or regional wall motion abnormality seen on echocardiography where other causes like coronary disease, sepsis, metabolic heart disease, congenital malformation could be ruled out and history concerning cardiomyopathy was negative.

Results

8 children – 5 girls and 3 boys – were admitted with the diagnosis of myocarditis (Table 1.). The median age of the patients was 1,16 years (range 0,25-8,71 years), the median of the follow-up time was 12,71 years (range 3,57-15,2 years).

We found an incidence rate of 1/10 000 of hospital discharges diagnosed with myocarditis at the Department of Pediatrics at the University of Debrecen, Hungary (1996-2010).

Clinical course

Three patients died of cardiogenic shock in the very acute phase of the disease (first, second and eighth day following admission). One patient subsequently developed left ventricular thrombus and cerebral embolization. The intracardiac thrombus resolved after heparin therapy and the child has mild residual right sided hemiparesis 12 years after the stroke. No serious complications occurred in the acute phase of the disease in 4 cases (Table2.).

Left ventricular function

The recovery of the severe initial left ventricular dysfunction (fractional shortening ≥ 30%) of the four survivors took 120 – 510 days (median 225 days). In the patient with perimyocarditis the regional wall motion abnormality disappeared on the 11th day of the disease.

None of the survivors developed dilated cardiomyopathy during the follow-up period.

Long term follow up and comorbidities

Patient 1 developed heart failure with elevated cardiac enzymes (aspartate-aminotransferase and alpha hydroxybutirate dehydrogenase) and LV dysfunction. Her cardiac function improved gradually and fractional shortening reached 30% after 120 days. Her LV size and systolic function remained normal during the next 15 years. From two years of age gradual visual impairment was detected. At the age of seven hypacusis was observed. She was also evaluated because of obesity and dyslipidaemia (cholesterin: 8,1 mmol/l, triglicerid 3,7 mmol/l). Impaired glucose tolerance (120' blood sugar 11,1 mmol/l), latent hypothyreosis (sTSH 4,6 mU/l, fT4 11,6 pmol/l) were confirmed.

At the age of 11 manifest type 2 diabetes mellitus was diagnosed (fasting blood sugar 17,5 mmol/l). Reviewing the symptoms, Alström syndrome was verified.

Patient 3, an 11-month-old boy, whose LV function recovered within 150 days was examined 8 years later because of abdominal pain and iron deficiency anaemia. Anti-endomysium antibody (EMA) positivity indicated duodenal biopsy, which revealed gluten sensitive enteropathy with subtotal atrophy (Stadium Mars IIIb). LV size and fractional shortening were normal when the diagnosis of coeliac disease was established. Gluten free diet resulted in the recovery of iron deficiency anaemia and the patient's growth accelerated.

Patient 4, an eight-year-old boy with perimyocarditis characterized by chest pain and very high troponin I level (115,9 µg/l) recovered within two weeks. A myocardium perfusion scintigraphy was performed, which revealed a negative result. Complete blood count was within the normal range (although MCV was 75 fl). Seven years later the symptoms of perimyocarditis recurred with high troponin T (4892 ng/l) and elevated pro-BNP levels (245 pmol/l). Coronary CT ruled out ischemic heart disease. The symptoms disappeared after three weeks. After recovery an EMA and tissue transglutaminase antibodies test was performed because of anaemia (HGB: 117 g/L, HTC: 0.37 MCV: 63.0 fL), which revealed a positive result. Duodenum biopsy confirmed the diagnosis of coeliac disease.

The normalization of the LV function in a 1-year-old infant (Patient 7) took 510 days. The persistent cough besides the improving trend of cardiac function drove us to search for another aetiology. Sweat test 40 days after the diagnosis of myocarditis proved to be positive and genetic evaluation revealed two heterozygote mutations of the CFTR gene (2184insA in 13a exon and deltaF508).

Table 1. Characteristics of patients on admission

Patient	Age (year)	Gender	Symptoms	Initial FS (%)	Low voltage on ECG	CK/TnI rise	Serology
1	0.58	Female	tachypnoea	7	Yes	Yes	
2	1.21	Male	cough, lack of appetite	10	No	No	
3	0.89	Male	cough, lack of appetite	10	No	No	
4	8.71	Male	chest pain	31	No	Yes	Influenza A
5	1.5	Female	diarrhoea, fatigue	19	No	Yes	Adeno-virus
6	1.45	Female	lack of appetite, fatigue	8	Yes	No	
7	1.11	Female	cough, lack of appetite, fatigue	6	Yes	Yes	
8	0.25	Female	tachypnoea, fatigue	13	Yes	Yes	

CK= creatine kinase, TnI= troponin I, FS= fractional shortening

Table 2. Long-term follow-up

Patient	Age (year)	Complication in the acute course	Time to recovery of LV function* (days)	Follow up duration (years)	Late outcome	Non-cardiac disease revealed during follow up
1	0.58	-	120	15.2	Complete cardiac remission	**Alström sy 8 yrs later**
2	1.21	LV thrombus, cerebral embolization	300	12.78	Complete cardiac remission Mild hemiparesis	-
3	0.89	-	150	12.71	Complete cardiac remission	**Celiac disease 9 yrs later**
4	8.71	perimyocarditis	11	8.32	7 yrs later recurrent perimyo-carditis	**Celiac disease 7 yrs later**
5	1.5	Death 2. day				
6	1.45	Death 3. day				
7	1.11		510	3.57	Complete cardiac remission	**Cystic fibrosis 1 month later**
8	0.25	Death 10. hour				

*Fractional shortening>30% or the cessation of wall motion abnormality

Conclusion:

To our knowledge this is the first pediatric study with a remarkably long follow-up focusing on co-morbidities associated with myocarditis. ***Myocarditis may precede the manifestations of other chronic pediatric diseases.*** Myocarditis or perimyocarditis may precede the clinical manifestation of celiac disease. The onset of severe cardiac dysfunction in infancy that recovers raises the possibility of myocarditis being the first symptom of Alström syndrome.