Introduction: acute myeloid leukemia (AML) is rare in children (incidence of 8 cases/million inhabitants/year). Extramedullary myocardial AML infiltration is an exceptional presentation defined as myeloid sarcoma (MS). Objective: to describe the first case in a 2 year old child.

Methods: a 2 year-old-child with a 2 week history of Bell paralysis plus eyes and lips edema was admitted for study. In physical examination no lymphadenopathy, organomegaly or skin pathology were found. There was testis inflammation and tachycardia. Blood count and cerebrospinal fluid (CSF) tests and cultures were taken, also bone marrow aspirate (BMA) and genetic tests. Extension study was performed.

Results: peripheral blood was abnormal: there was neutrophilia with circulating myelocytes, 24% of blast cells in blood count, normal platelets and LDH 980 and no anemia (hemoglobin 15mg/dl). X-ray revealed cardiomegaly, abdominal echography showed discrete hepatomegaly and mild renal hydronephrosis. Scrotal echography suggested tumoral infiltration. BMA diagnosed AML M4/M5 (40% blasts). CSF proved infiltration with hypercelularity and 97% blasts. Echocardiography revealed a diffuse and heterogeneous cardiac infiltration, involving pericardium and myocardium associated with a moderate circumferential pericardial effusion without hemodynamic compromise and preserved cardiac function. 3 days after induction chemotherapy treatment hemodynamic instability started with increased global infiltration and pericarditis. IV furosemide and milrinone were used with good results. Control echocardiography 3 weeks after (patient in first complete hematological remission) showed a significant global decrease in the cardiac infiltration with a minimal residual pericardial effusion, but showed a persistent decrease in left ventricular ejection fraction (LVEF=40%). The pretransplant echocardiographic control, after two cycles of chemotherapy treatment, showed recovery of systolic and diastolic biventricular function, without cardiac infiltration or pericardial effusion.

Conclusions: The difficulty diagnosing MS requires a high degree of clinical suspicion in the setting of unusual leukemic presentations. AML is an exceptional cause of myocardial infiltration. Prompt recognition of the underlying leukemia and initiation of appropriate therapy are key to reducing overall morbidity and mortality. Echocardiographic aided both diagnosis and assessment of response to treatment of the cardiac infiltration.