NAKNADNO PRISTIGLI SAŽECI | LATE SUBMITTED ABSTRACTS

Spužvasta kardiomiopatija

Noncompaction cardiomiopathy

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Sažetak

Izolirani nekompaktni lijevi ventrikul ili tzv. "spužvasta kardiomiopatija" (engl. *isolated left ventricular non-compaction — ILVNC*) rijetka je bolest, po Američkom kardiološkom društvu klasificirana kao primarno nasljedna, a prema Europskom kardiološkom društvu kao neklasificirana kardiomiopatija. Karakterizirana je promijenjenom morfologijom miokarda lijeve klijetke (LV) najčešće inferopaikalno i apikolateralno. Zahvaćeni miokard ima kompaktni i nekompaktni sloj, izrazito je trabekuiliran, s brojnim dubokim međutrabekulskim recesusima koji komuniciraju sa šupljinom LV. Promijenjena morfologija miokarda posljedica je poremećene intrauterine morfogeneze endomiokarda. Izolirani oblik bolesti javlja se uglavnom kod odraslih, a u djece je bolest obično udružena s drugim prirođenim srčanim manama i malformacijama uključujući hidrops fetusa i sindrom iznenadne smrti novorođenčadi. Poremećena morfogeneza miokarda predisponira razvoju dilatacije, hipertrofije i disfunkcije. Genetski poremećaj se povezuje s mutacijama proteina mitohondrija, citoskeleta, Z-linije i sarkomere. Bolest može biti bez simptoma, ili se očituje atrijskim i/ili ventrikulskim aritmijama, zatajivanjem srca, tromboembolijama ili iznenadnom smrću. Prevalencija bolesti u općoj populaciji kao i prirodni tijek nisu dobro poznati. Nekompaktni LV opažen je i u rođaka bolesnika s hipertrofijskom i dilatacijskom kardiomiopatijom, ali i u dijela zdravih osoba. Međutim, ne treba poistovjećivati hipertrabekulaciju LV s razvijenim oblikom "spužvaste" kardiomiopatije.

Dijagnoza se postavlja prvenstveno ehokardiografijom, ali također i lijevom ventrikulografijom, kompjutoriziranom tomografijom i nuklearnom magnetskom rezonancijom.

Diferencijalna dijagnoza uključuje dilatacijsku kardiomiopatiju, hipertenzivnu bolest srca, apikalni oblik hipertrofijske kardiomioipatije i endomiokardnu fibrozu. Ehokardiogram je abnormalan u svih bolesnika. Preporučeni dijagnostički kriteriji temelje se na opažanjima China, Jennia i Stollbergera. Chin je na temelju opažanja u osam bolesnika uzeo kao kriterij odnos udaljenosti od epikarda do recesusa (x) i od epikarda do vrha trabekulacije (y): x/y

Abstract

The isolated left ventricular non-compaction (ILVNC) is a rare disease. According to the American Heart Association, classified as a primary congenital disease, and according to the European Society of Cardiology it is designated as non-classified cardiomyopathy. It is characterized by unchanged left ventricular (LV) myocardial morphology, usually inferoapically and apicolaterally. The affected myocardium has a compact and non-compact layer, very trabeculated with a great number of deep intertrabecular resources that communicate with the LV chamber. The changed myocardial morphology is the consequence of disturbed intrauterine endomyocardial morphogenesis. The isolated form of the disease occurs mainly in adults but in children the disease is usually accompanied with some other congenital heart defects and malformations, including the fetal hydrops and newborn sudden death syndrome. The disturbed myocardial morphogenesis makes patients inclined to development of dilatation, hypertrophy and dysfunction. Genetic disorder is connected with mutation of mitochondrions, cytoskeletons, Z-line and sarcomeres. The disease may be with no symptoms or is characterized by atrial and/or ventricular arrhythmias, heart failure, thromboembolism or sudden death. The disease prevalence in general population and the natural course are not well known. Non-compact LV is perceived in a patient's cousin with hypertrophic and dilatative cardiomyopathy, but also in one segment of healthy persons. However, we should not equalize the LV hypertrabeculation with developed form of "non-compaction" cardiomyopathy.

The diagnosis is made mainly by echocardiography, but also by the left ventriculography, computerized tomography and nuclear magnetic resonance.

The differential diagnosis includes dilatative cardiomyopathy, hypertensive heart disease, apical form of hypetrophic cardiomyopathy and endomyocardial fibrosis. The echocardiogram is abnormal in all patients. The recommended diagnostic criteria are based on perceptions of Chin, Jenni and Stollberger. Chin has, based on perceptions in 8 patients, used a criterion of ratio of distance from the epicardium to recesus (x) and from the epicardium to

jednako ili manje od 0,5 ukazuje na prisutnost spužvaste kardiomiopatije. Jenni i kolege preporučili su sljedeće kriterije (pregled u kratkoj parasternalnoj osi): 1. odsustvo drugih strukturnih abnormalnosti srca, 2. izrazito zadebljana stjenka lijeve klijetke koja se sastoji od dva sloja — tankog epikardnog koji je kompaktni i debelog, endokardnog sloja s izraženom trabekulacijom i dubokim recesusima (odnos nekompaktnog prema kompaktnom sloju veći od 2:1); 3. obojenim doplerom dokazan protok unutar recesusa, i 4. izrazita trabekulacija apikalno, inferoapikalno i/ili apikolateralno. Često je prisutna hipokinezija nekompaktnih segmenata. Druge ehokardiografske abnormalnosti uključuju globalnu hipokontraktilnost, dijastoličku disfunkciju, nalaz muralnih tromba i abnormalnu strukturu paiparnih mišića.

Prognoza bolesti nije dovoljno poznata; petogodišnje preživljavanje je oko 60%.

Liječenje je medikamentno uz eventuanu ugradnju kardioverter defibrilatora, a u bolesnika s refraktornim zatajivanjem srca indicirana je transplantacija.

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the tip of the trabeculation (y): x/y equal or less than 0,5 indicates non-compaction cardiomyopathy. Jenni and the colleagues recommended the following criteria (examination in short parasternal axis): 1. absence of some other structural cardiac abnormalities, 2. extremely thickened LV wall consisting of the two layers — a thin epicardial layer that is compact and of thick, endocardial layer with strong trabeculation and deep recuses (that is, non-compact towards compact layer greater than 2:1); 3. a proved flow within recesuses by using color Doppler, and 4. an extreme trabeculation apically, inferoapically and/or apicolaterally. The hypokinesia of non-compaction segments is frequently present. Some other echocardiographic abnormalities include global hypocontractility, diastolic dysfunction, the findings of mural thrombi and abnormal papillary muscle structure.

The prognosis for diseases in not sufficiently known, a five-year survival is around 60%.

The treatment is envisaged with drugs with a possibility of implantation of defibrillator cardioverter thereby indicating transplantation in patients with refractory heart failure.