Brugadin sindrom: implantabilni kardioverter-defibrilator svima? Prikaz slučaja

Brugada syndrome: an implantable cardioverter defibrillator for everyone? Case report

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Brugadin sindrom je kanalopatija koja se nasljeđuje auto-
somno dominantnim obrascem, a karakterizira ju izosta-
nak strukturne bolesti srca, karakterističan elektrokardi-
ografski zapis te povećana incidencija iznenadne srčane
smrti. Poremećaj se povezuje s mutacijom gena SCN5A
koji kodira a jedinicu natrijskog kanala. Prevalencija bole-
sti se procjenjuje na 5/10.000, a zaslužna je za 20% iznenad-
nih srčanih smrti u bolesnika bez strukturne bolesti srca
te 4-12% svih iznenadnih srčanih smrti u općoj populaciji.
Prema Međunarodnom registru Brugadinog sindroma 25%
pacijentata sa ovom bolesti tijekom svog života doživi
ili iznenadnu srčanu smrt ili ventrikularnu fibrilaciju sa
srednjom dobi od 42 godine. Abortirana iznenadna srčana
smrt dogodi se u 7,7% pacijenata, 1,9% u pacijenata koji su
imali sinkopu, a tek u 0,5% asimptomatskih pacijenata. Je-
dini djelotvorni modalitet liječenja i prevencije iznenadne
srčane smrti je ugradnja implantabilnog kardioverter-de-
fibrilatora, što je rezervirano kod svih simptomatskih bole-
snika. Kod asimptomatskih pacijenata stratifikacija rizika
je neobično važna uzevši u obzir visoku cijenu terapije te
ozbiljnost ishoda bolesti.

U sklopu rada bit će prikazan slučaj 64-godišnjeg asimp-
tomatskog bolesnika, bez obiteljske anamneze iznenadne
srčane smrti, u kojeg je ajmalinskim testom dokazano po-
stojanje Brugadinog sindroma.

Brugada syndrome is a channelopathy inherited as an
autosomal dominant trait and characterized by the lack
of structural heart disease, specific electrocardiographic
record and an increased incidence of sudden cardiac
death. This disorder has been linked with mutations in
genes encoding the a unit of SCN5A sodium channel. The
prevalence of the disease is estimated to be 5 in 10.000
people, and is considered to be responsible for 20% of
sudden cardiac death in people without structural heart
disease and 4-12% of all sudden cardiac deaths in the
general population. According to the International Regis-
try for Brugada syndrome, 25% of patients experience ei-
ther sudden cardiac death or ventricular fibrillation dur-
ing their life (a mean age being 42 years). Aborted sudden
cardiac death occurs in 7.7% of all the patients, 1.9% of the
patients with syncope, and in only 0.5% of the asymp-
tomatic patients. The only effective treatment and preven-
tion of sudden cardiac death is the implantation of an
implantable cardioverter defibrillator, which is reserved
for all the symptomatic patients. In asymptomatic pa-
tients, considering the high cost of treatment and sever-
ity of illness, risk stratification is extremely important.

A case of 64-year-old asymptomatic patient, with no
family history of sudden cardiac death, diagnosed with
Brugada syndrome using the ajmaline test, will be pre-
sented.

LITERATURE

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