ICD Implantation In Patient with Cardiac Amyloidosis
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Clinical Question
In a 65-year-old male with cardiac involvement of light chain (AL type) amyloidosis, will an implantable cardioverter-defibrillator prevent episodes of sudden cardiac death?

Case Scenario
- 65 year white male with dyspnea, fatigue, angina, is evaluated following a syncopal episode
- Physical exam demonstrates the presence of jugular venous distension, S3 gallop, bilateral basal rales, and lower extremity edema
- Echocardiogram: ejection fraction 52%
- EKG: low voltage complexes
- Endomyocardial biopsy: light chain amyloid deposits.
- Medications: furosemide, metoprolol; melphalan

Search Strategy
- Pub Med: Clinical Queries search tool, Therapy Category, Broad Scope
- Search Terms: “cardiac amyloidosis’ AND “cardioverter defibrillator”
- Search yielded three references with one applicable to the topic
- Pub Med: Advanced Search with Search Terms “cardiac amyloidosis” AND “cardioverter defibrillator”
- Search yielded 19 references with 4 appropriate for the topic
- Clinical Key: Search with terms “cardiac amyloidosis” AND “cardioverter defibrillator” with meta-analysis filter
- Search did not yield any studies that applied to the topic

Table 1-Successful Defibrillation of ICD in patients with AL Type Cardiac Amyloidosis

<table>
<thead>
<tr>
<th>Reference</th>
<th>Citation</th>
<th>LOE</th>
<th>Population</th>
<th>N</th>
<th>Successful Termination of VT/VF</th>
<th>Successful Defibrillation (95% CI)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Kristen et al. (2008) 4</td>
<td>AL Type Amyloidosis with ICD Implantation</td>
<td>2</td>
<td>2</td>
<td>100%</td>
<td>(29%, 100%)</td>
<td></td>
</tr>
<tr>
<td>Karin et al. (2013) 4</td>
<td>AL Type Amyloidosis with ICD Implantation</td>
<td>10</td>
<td>7</td>
<td>70%</td>
<td>(39%, 90%)</td>
<td></td>
</tr>
<tr>
<td>Lin et al. (2013) 4</td>
<td>AL Type Amyloidosis with ICD Implantation</td>
<td>15</td>
<td>12</td>
<td>80%</td>
<td>(54%, 94%)</td>
<td></td>
</tr>
<tr>
<td>Varr et al. (2014) 4</td>
<td>AL Type Amyloidosis with ICD Implantation</td>
<td>5</td>
<td>4</td>
<td>80%</td>
<td>(36%, 98%)</td>
<td></td>
</tr>
</tbody>
</table>

Table 2-Long Term Outcome for AL Type Cardiac Amyloidosis patients with ICD Implantation

<table>
<thead>
<tr>
<th>Reference</th>
<th>Citation</th>
<th>LOE</th>
<th>Population</th>
<th>N</th>
<th>Follow-Up</th>
<th>Survival Rate (alive; deceased)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Kristen et al. (2008) 4</td>
<td>AL Type Amyloidosis with ICD Implantation</td>
<td>19</td>
<td>28 Months</td>
<td>52%</td>
<td>(10; 9)</td>
<td></td>
</tr>
<tr>
<td>Karin et al. (2013) 4</td>
<td>AL Type Amyloidosis with ICD Implantation</td>
<td>10</td>
<td>28 Months</td>
<td>50%</td>
<td>(5; 5)</td>
<td></td>
</tr>
<tr>
<td>Lin et al. (2013) 4</td>
<td>AL Type Amyloidosis with ICD Implantation</td>
<td>33</td>
<td>24 Months</td>
<td>30%</td>
<td>(10; 23)</td>
<td></td>
</tr>
<tr>
<td>Varr et al. (2014) 4</td>
<td>AL Type Amyloidosis with ICD Implantation</td>
<td>15</td>
<td>18 Months</td>
<td>40%*</td>
<td>(2; 3)</td>
<td></td>
</tr>
</tbody>
</table>

Background
- Amyloidosis is an idiopathic disease process in which abnormal antibodies build up due to excessive proliferation in bone marrow. The antibodies build up and form amyloid fibrils that deposit into multiple areas of the body.
- When the buildup occurs in the heart, patients often go into heart failure, predominantly right sided.
- Patients suffer from life-threatening ventricular arrhythmias. There is high risk for sudden death and death due to progressive cardiomyopathy. Average survival once diagnosed with cardiac amyloidosis: less than one year.
- Guidelines do not recommend invasive procedure of implantation of defibrillator due to the fact that life expectancy is so low.

Conclusions
- There is not enough evidence to determine whether or not the implantation of a cardioverter-defibrillator will be beneficial for the patient in this clinical scenario.
- There is clear evidence that the ICD implantation prevents sudden cardiac death in the small subset patients that suffer from tachyarrrythmias.

Application to Patient
- It is unknown whether or not an implantable cardioverter-defibrillator will be beneficial in this patient.
- More information must be obtained to determine whether or not the patient suffers from any tachyarrrythmias.

Future Directions
- In the future it would be beneficial to first develop a case-control study or other higher level of evidence that addresses this topic.
- Rarity of cardiac involvement of AL type amyloidosis may not ever make it possible to develop a randomized controlled trial.
- If researchers are able to determine specifically which patients will suffer from tachyarrrythmias then we will be able to direct treatment towards those patients.

References
5) “Cardiac amyloidosis very high mag he” by Nepron - Own work. Licensed under CC BY-SA 3.0 via Commons - https://commons.wikimedia.org/wiki/File:Cardiac_amyloidosis_very_high_mag_he.jpg?uselang=en

Cardiac amyloidosis (lighter pink infiltrate at top)