PROPHYLACTIC PACemaker IMPLANTATION IN FAMILIAL AMYLOID POLYNEUROPATHy
WITH MINOR CONDUCTION DISORDERS.

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Background:
Familial amyloid polyneuropathy (FAP) is a dominantly inherited neuromuscular disease caused by mutated transthyretin. FAP cardiopathy is characterized by cardiac infiltration leading to conduction disorders, with increased risk of sudden death. Prophylactic cardiac pacing may be considered in asymptomatic patients exhibiting minor conduction disorders. However, the potential benefits remain to be documented.

Objective: The hypothesis that prophylactic pacemaker implantation may protect FAP patients from bradycardia due to transient or permanent AV block was tested.

Methods:
From January 1999 to January 2010, fifty-one patients with genetically proven FAP (53 ± 13 years old) and minor conduction disorders were implanted with prophylactic dual chamber cardiac pacemaker(cf. Figure 1&2). Both surface ECG and the complete PM interrogation including temporary pacemaker inhibition and download of the device memory collected at each follow-up visit were retrospectively analyzed over a mean observation period of 43 ± 35 months.

Results:
Before pacemaker implantation, ECG was abnormal in 40/51 patients (78%), consisting of intra ventricular conduction disorders (n=20), first degree AV block (n=11) or both (n=9). In the remaining 11 patients, HV interval ranged from 50 to 92 ms (cf. table 1). During follow-up, temporary pacemaker inhibition indicated pacemaker dependency with high-degree AV block in 13/51 patients (25%) (cf. Figure 3, 4 & 5). The time-to-dependency upon pacemaker was 66 ± 42 months (range 5-130).

Conclusion:
In FAP with minor conduction disorders, prophylactic pacemaker implantation prevented major cardiac events in 25% patients over a 43 months mean follow-up. It is suggested that prophylactic PM implantation prevented symptomatic bradycardia in these patients.