THE MANAGEMENT OF PROXIMAL AORTA IN MARFAN SYNDROME
PhD Thesis Summary

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The purposes of proper investigations are retrospective clinical analyses and evaluation of medical history, prehospital and perioperative management in Marfan syndrome's condition proximal aorta manifestations.

The database comprises two decades (1988-2010) period in a representative surgical center. Observational casuistics of the Cardiovascular Clinics of Semmelweis University, Budapest on performed mandatory prophylactic and emergency aortic operations.

The database comprises - according to objective's criteria -data of 76 operated patients, the medical history from data sheets, surgery protocols, emission documents and protocol related polling and processing.

- The diagnosed or undiagnosed prehospital period; the preoperative period, the emergency and timing strategy; the intraoperative guidelines in aortic dilatation and dissection of proximal aorta, aortic regurgitation of Marfan patients; the postoperative 30 days period management, outcomes
- The interpretative comparison of emergency and preventive surgical management and conclusive deductions based on best BETS method evaluation

The sketch of these structure in two parts, the general aspects and the proper observations, contains three, respectively seven chapters:

- General parts (3 chapters, 69 pages): revised literature of Marfan syndrome and proximal aortic manifestations, fundamental (etiopathogenesis, surgical anatomy), clinical features (classification, diagnostic criteria, preventive care, pharmacotherapy) and surgical strategy
- The personal observations (7 chapters, 152 pages) evaluate according to the aims of the demographic and clinic patient's casuistics, the protocol and methodology, in following main proximal aortic complications:
  - Acute and chronic aortic dissections (16/76; 11/76 patients)
  - Aortic regurgitation, annulo-aortic ectasia in MFS (30/76 patients)
  - The preventive operations in ascendent aorta dilatation (19/76 patients)
- Our study includes two associated chapters:
  - The fibroarchitecture of aortic root valvular supporting structure (Chapter V.)
  - The neuroprotectivity of axillary selective cerebral perfusion in cardiopulmonary bypass
- Adnotation regarding the aortic surgery revision in MFS
- Statistical analyses of gained numerical data in proximal aortic manifestations of MFS

1. The aortic root-valve correlative pathology and surgery of their functional assonance with cardiac cycle and hemodynamical forces asserted by an immuno-histological study of structural interrelations regarding the dynamic adhesions of root-valve connections. The supporting structures are fibroelastic and mechano-architectural supported systems, transferring the loads of leaflets to aortic elastic layers, including through compliance/impedance adaptivity resulting in potential energy storage.

2. Diagnosis of MFS was done with various clinical examinations, as there was no quick test available for genetic examinations. The diagnosis was established by help of the etiological genetic factors such as direct familial anamnensis, the Marfan body habitude and build-up, the young age and the Ghent
nosological criteria based on the six organ-system manifestations including the most important pathognomic aortic root dilatation.

3. Cannulation methods of extracorporeal circulation in surgery of proximal aortic manifestations in MFS are key conditions, regarding the advantages and detriments of different sites for the cannula and perfusion. In the preventive and elective surgery of aortic dilatation and annulo-aortic ectasia, the aorto-atrial or aorto-bicaval cannulations were used preferentially. Two cohorts, comparing the anterograde cerebral perfusion providing axillary and retrograde aortic flow causing femoral arterial cannulations, it is to conclude that the mortality rate and neurological mortality and morbidity (stroke) decreased with 50%, the latter respectively with 80% in favor for axillary cannulations – a great technique in avoiding the embolism and mal- or hypoperfusion of the central nervous system (brain).

4. The hypothermic protection associated with cannulation techniques is the most important procedure to prevent or reduce the ischemic damage of the brain – not mentioning the heart - in proximal aortic operations. In the observational casuistics the degree of hypothermia used is differentiated correlation with cannulation methods, surgery types. Under circumstances of aortic cannulation techniques preferentially in prophylactic surgery, mild hypothermia was applied (32-30°C), in axillary and femoral artery cannulations, the hypothermic conditions were moderate (28-22°C) or deep (20-14°C).

5. The proximal aortic manifestations operative procedures are mandatory, the strategical paradigms privilege the preventive surgery under MFS condition. In different surgical treatment modalities (preventive electve, urgent, emergency) the paramount method for aortic ascendent reconstruction is the application of conduit prosthesis with valve implanted according to Bentall-DeBono. In 79% of aortic root reconstruction cases (60/76) the classic techniques were applied with different types of reimplantation of coronary artery orifices into the Dacron graft, the crucial maneuver intraoperatively outlined usually with the use of different techniques (direct, Button, Cabrol, SVG-CABG). Combining the use of Bentall method for one staged repair of aortic root with associated arch reconstruction or mitral insufficiency surgery has fundamental significance. (20%, 15/76)

Use of the David procedure, a valve sparing alternative, in reconstruction of MFS affected aortic root only at specific indication premises is considerable (pregnancy), namely the mild durability of the native valvules is time limited due to the progressive nature of the MFS disorder on biomechanical and structural properties. (Valve sparing reimplantation: 4%, 3/76)

6. Regarding the 30 post-operative days outcome at the cohorts, the mortality was a total 9%, 7 patients of 76 exited at proximal aortic operations due to following reasons in details: In the acute AAD group there were 4 deaths (25%) due to ischemic stroke, cerebral hypoxia – 3 cases, myocardial infarct due to dissection of coronary artery ostium – 1 case. In the proximal ChAD group, there were 2 exits (18%): 1 case because of hypoperfusion syndrome and another case due to depression and severe negativism (these two operations were Bentall procedure+ arch surgery performed). In the annulo-aortic ectasia group one patients exited (3%) due to coronary artery ostium thrombosis. At the preventive surgery indication group -due to aortic dilatations - there were no operative mortality in the first 30 days after surgery.

7. The survival of all patients operated with proximal aortic manifestations was of high rate, as the exemplification supported with results at 5 years after emergency acute aortic dissection dissection and the prophylactic cohort group operated. The five years survival in preventive proximal aortic surgery is almost 50% better then acute AAD at MFS patients.

8. The final inductive inferences approaching the MFS proximal aortic manifestations in a retrospective viewpoint rises out of the following:

- The early screening and diagnosis of MFS and it's aortic manifestations
- The increased role of the National Marfan Foundation network and the draft of a national register for proximal aortic manifestations' treatments and demographics data
- The life long management and care of MFS patients, the prevention and previsibility of aortic manifestations and the prognostic course of the disease
- Helping the individual, in the framework of an optimal medico-psycho-social approach to achieve and hold trust in expectancies about quality and extent of life besides the surgical management.