ROLE OF SPECIALIZED CENTRE AND TEAMWORK IN THE DIAGNOSIS OF MARFAN SYNDROME AND PREVENTION OF ACUTE AORTIC DISSECTION

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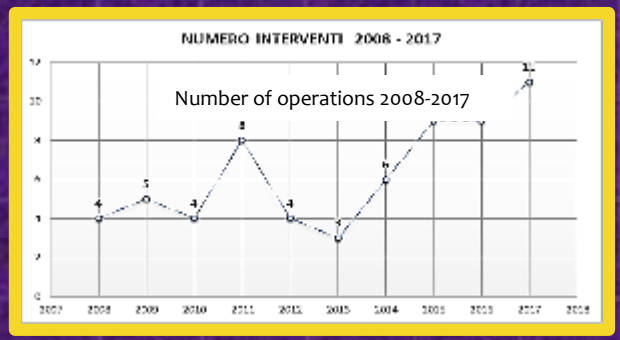
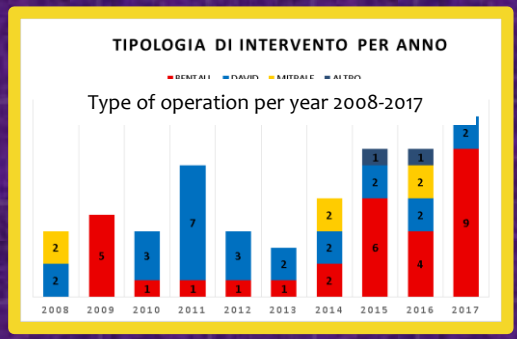
Summary: People with Marfan Syndrome had an increasing risk of aortic dissection related to the quality of the aortic wall. Current guidelines indicated an aortic diameter ≥ 50 mm as good parameter for surgical indications. However recent studies showed that most of dissected aorta had a diameter < 50 mm. In our opinion the diameter is not enough, other factors need to be considered in order to prevent aortic dissection, such as the morphology of the aortic root, the prolapse of Valsalva sinus and other genetic and clinical aspects. In our Hospital we established a specialized Centre to improve Marfan Syndrome diagnosis and optimize surgical indication in order to prevent catastrophic complications. :

Materials and Methods:We enrolled patients admitted to the Marfan Syndrome Centre of Tor Vergata University from March 2008 to December 2017. These patients have been evaluated by a teamwork composed by different figures: cardiologists, cardiac surgeons, oculists, orthopaedics, dentists, genetics and nurses. Marfan Syndrome diagnosis has been established, clinically, according to the Ghent's Criteria. Patients have been followed periodically (every six months, every year) according to the severity of the pathology. An aortic root diameter above 45 mm, associated to particular morphology of aortic root or Valsalva sinus prolapse with or without aortic regurgitation was used as good criteria for surgical indication.

Results: From 1323 patients admitted, we enrolled a total of 460 patients. Of these 260 patients had Marfan Syndrome, 10 patients had Loeys-Dietz Syndrome and the others 190 were Marfan like. Of these enrolled patients, 95 underwent cardiac surgery. In particular 47% had a Bentall-DeBono Operation, 40% a David Operation and 13% other operations. Related to the great and intensive activity of the teamwork and, at the same time, to the careful follow-up of these patients an increasing number of surgical operations has been done in the years. Accordingly we noticed a reducing incidence of acute aortic dissection and sudden deaths in our patients





Conclusions: A specialized Centre of Rare Disease is very important in the management of Marfan's patients in order to improve the diagnosis and optimize surgical indications. A good surgical timing is necessary to prevent catastrophic complication like acute aortic dissection and rupture. The diameter of the aortic root is not enough for surgical indications. In our opinion the morphology of the aortic root and the prolapse of Valsalva sinus, associated to clinical and genetic aspect should be good predictors of aortic dissection in Marfan's patients

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