

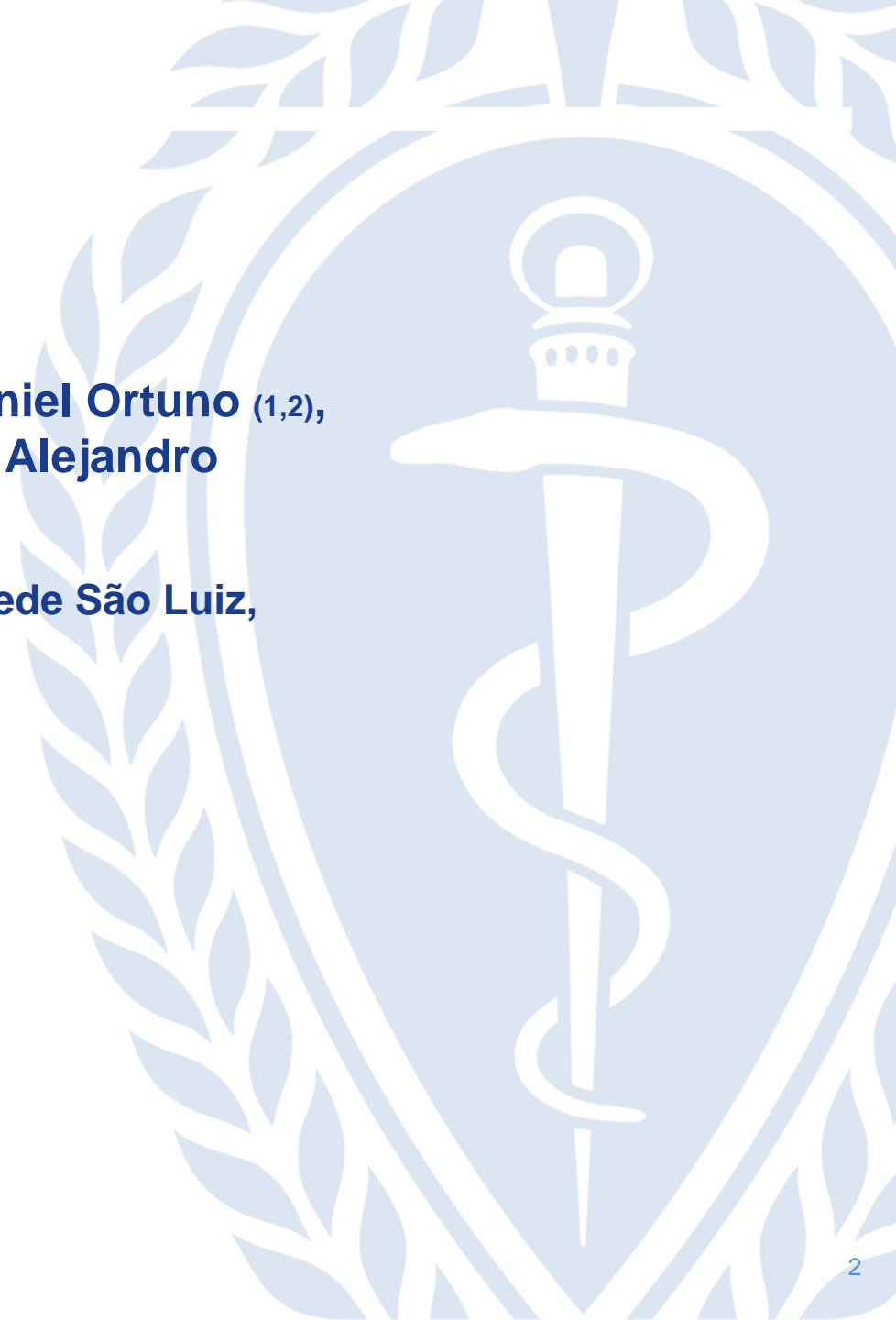
A Rare Case of Ascending Aortic Aneurysm in a Pediatric Patient

Analysis and Management of One Patient with Cowden Syndrome

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Presentation

- male patient at 2 year of age
- shortness of breath and failure to thrive
- severe aortic regurgitation
- heart failure



- **large aneurysm of the aortic root and ascending aorta**
- **aortic valve was deemed tri-valvular**
- **not possible to compensate the patient and surgical intervention was recommended.**

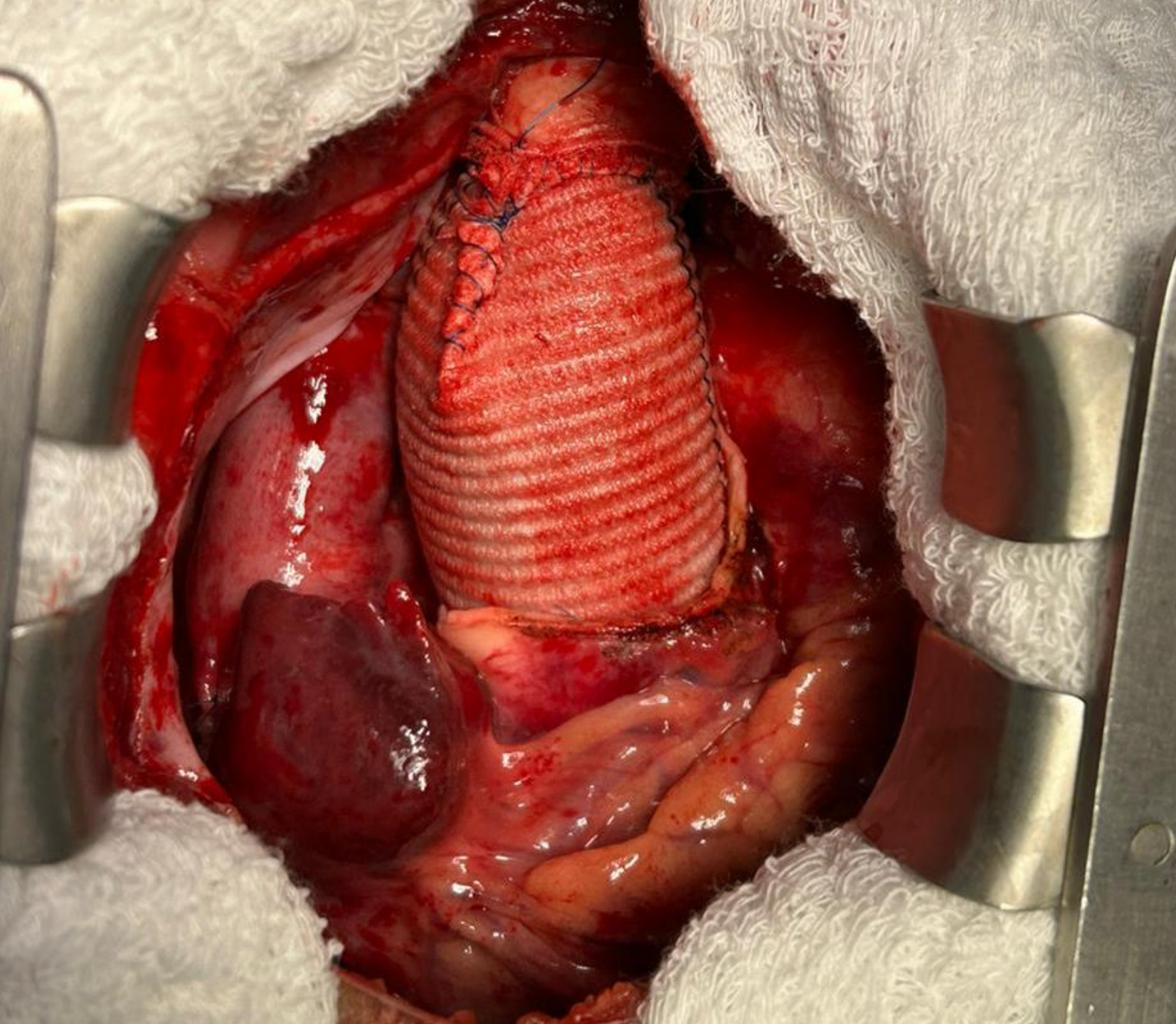




- **Bentall-de Bonno operation**
- **biological pericardial prostheses INSPIRIS number 23 (Edwards LifeSciences), mounted on a 30 mm dacron graft**
- **uneventful - extubated the following day**

- **post operative day 4 - surgical re-exploration due to cardiac tamponade**
- **discharged home on post operative day 14**
- **Late follow-up at 6 months shows good surgical result.**





Comments

- **Cowden Syndrome, mutation on the PTEN gene (phosphatase and tensin homologue).**
- **PTEN mutations are related to vascular anomalies**
- **no literature correlated this finding with aneurysm of the aorta**
- **limited number of cases, not possible to draw more accurate conclusions**



Conclusion

- **In this particular case it was possible to implant an adult-size prostheses to ensure adequate growth to the patient**
- **for a future intervention bioprosthesis will be replaced with a mechanical one.**

