Open Thoracic and Thoraco-abdominal Aortic Repair in Patients with Connective Tissue Disease
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OBJECTIVE
To present current results of open complex aortic repair in patients with connective tissue disease (CTD).

METHODS
Retrospective cross-border, single centre study
February 2000 to April 2016
72 aortic operations on 65 patients with CTD
41 male, median age 41 years [range 19–70 years]
56 patients (86%) with previous aortic repair (71 open, 4 endovascular)
33 patients (51%) operated before at the site of the procedure reported here
Procedures:
8 emergency operations (11%)
- Aortic arch revision (n = 1; 1%)
- Descending thoracic aortic repair (n = 11; 15%)
- TAAA type I repair (n = 12; 17%)
- Type II repair (n = 29; 40%)
- Type III repair (n = 12; 17%)
- Type IV repair (n = 5; 7%).
- Ascending aorta and/or the aortic arch (n = 2; 3%) and (n = 8; 11%)
7 patients (10%) underwent staged procedures
Median follow-up: 42 months (0.5–180 months)

RESULTS
In hospital mortality: 14% (n = 9)
- Haemorrhage 3/9
- Neurological 3/9
- Cardiac 2/9
- Pulmonary 1/9
Paraplegia/paraparesis: 2%/5%
Temporary dialysis: 11%
Myocardial infarction: 3%
Stroke: 2%
Freedom from re-intervention 85%
1 year survival 80%
Overall survival 75%

CONCLUSIONS
Open TAA(A) repair is a durable therapy for patients with CTD. Often being performed as revision surgery, it can be associated with relevant risks and should therefore be reserved for specialised centres. Staged procedures and thus reducing operating time, if applicable, should be preferred.