

Endovascular Thoracoabdominal Aneurysm Repair in Patients with Connective Disease

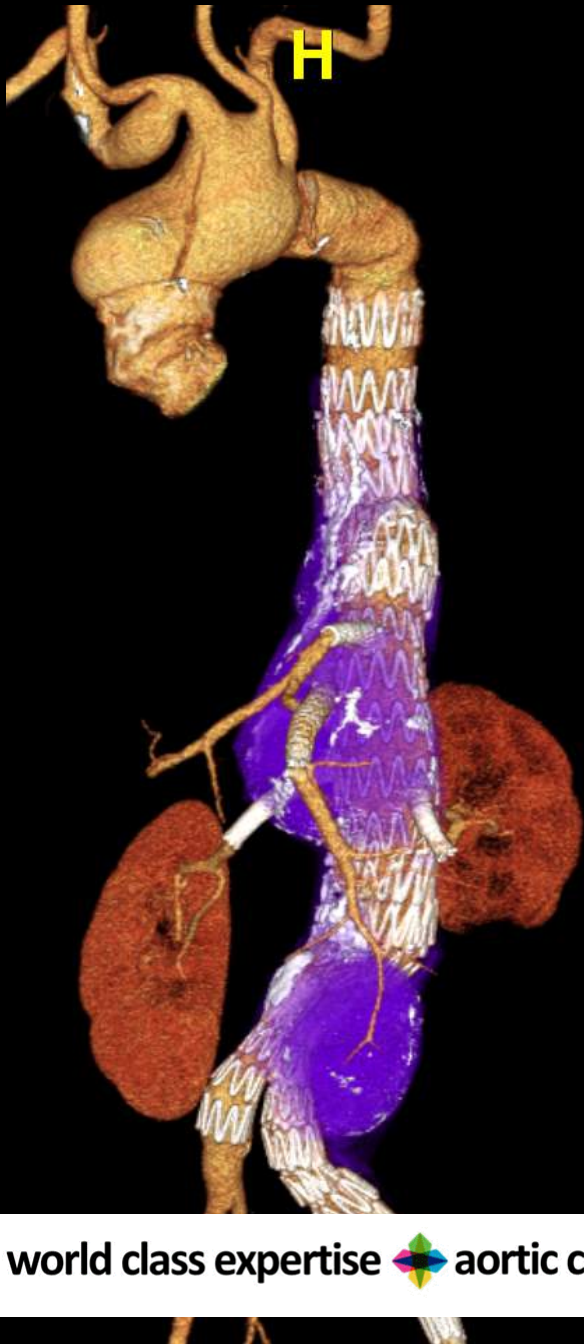
world class expertise  aortic care

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Royal Free London

Pertinent Disclosures

- CYDAR Medical: Research Collaboration and Consultation
- Siemens Imaging: Research Collaboration
- Cultivate Labs: Research Collaboration
- Cook Medical: Educational Grant and Proctoring

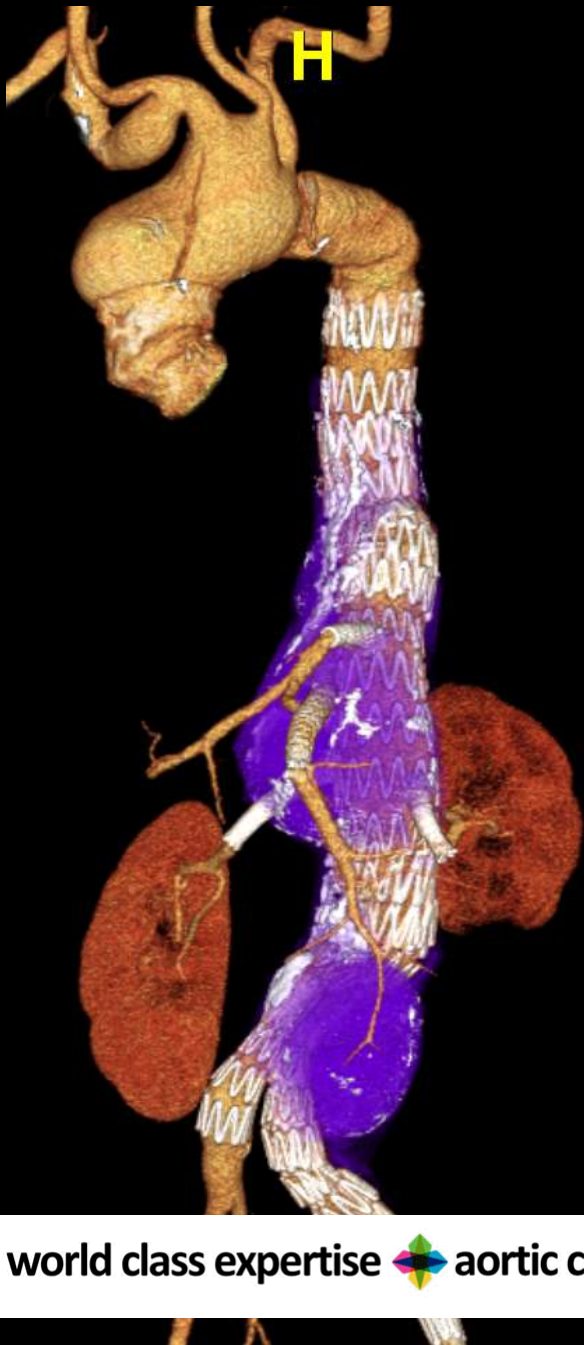
The **Royal Free Aortic Charity** Funds
Research and Educational Activities to
support the Aortic Team at Royal Free
Foundation Trust



**What Constitutes
CTD?**

What do we know?

**Are there Any Rules
in CTD?**



What Constitutes CTD?

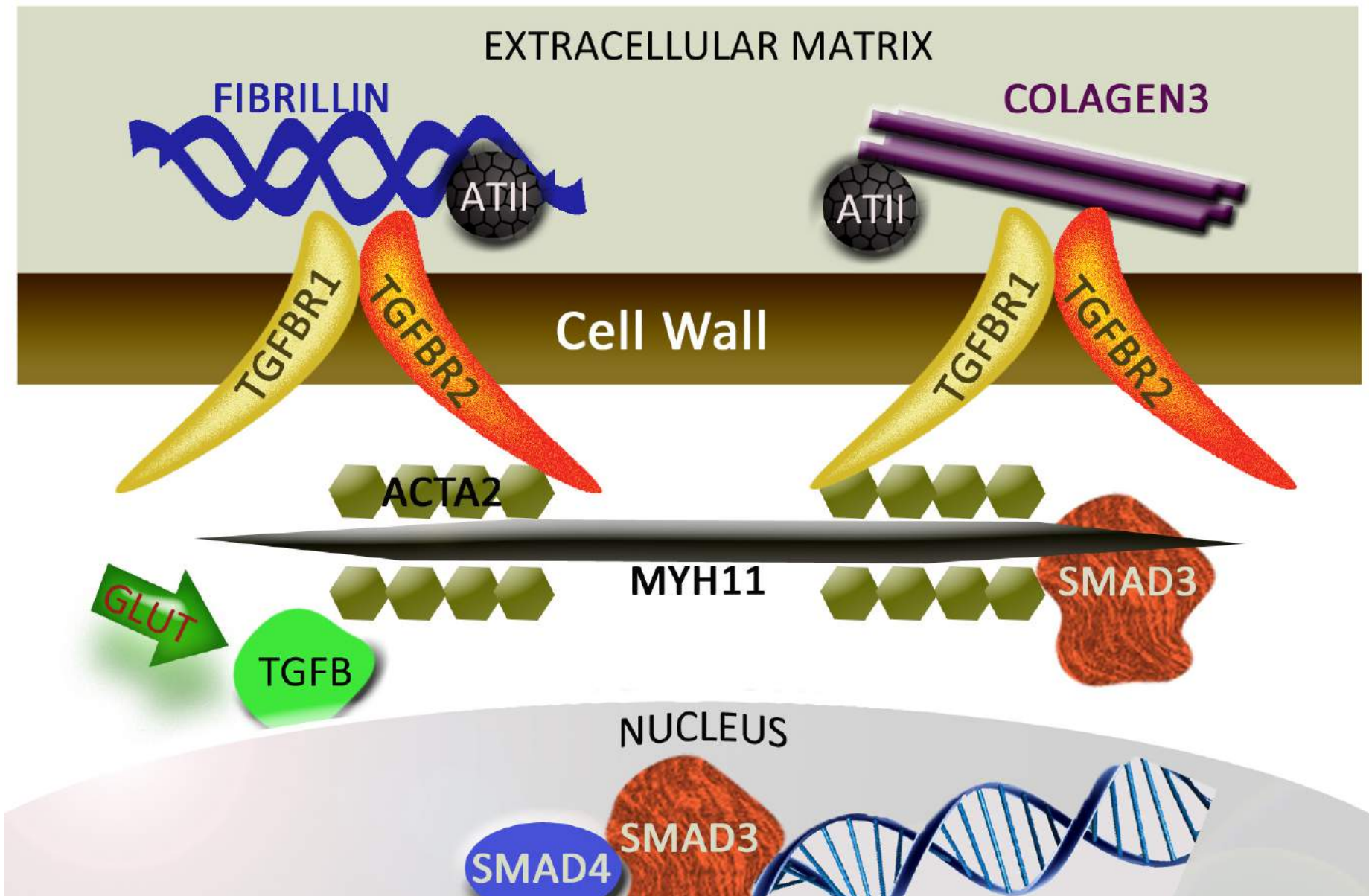
A

H

P

F





Marfan Syndrome

Ectopic Lentis
Overgrowth
Pectus
Scoliosis
Aortic Root

Ehlers Danlos

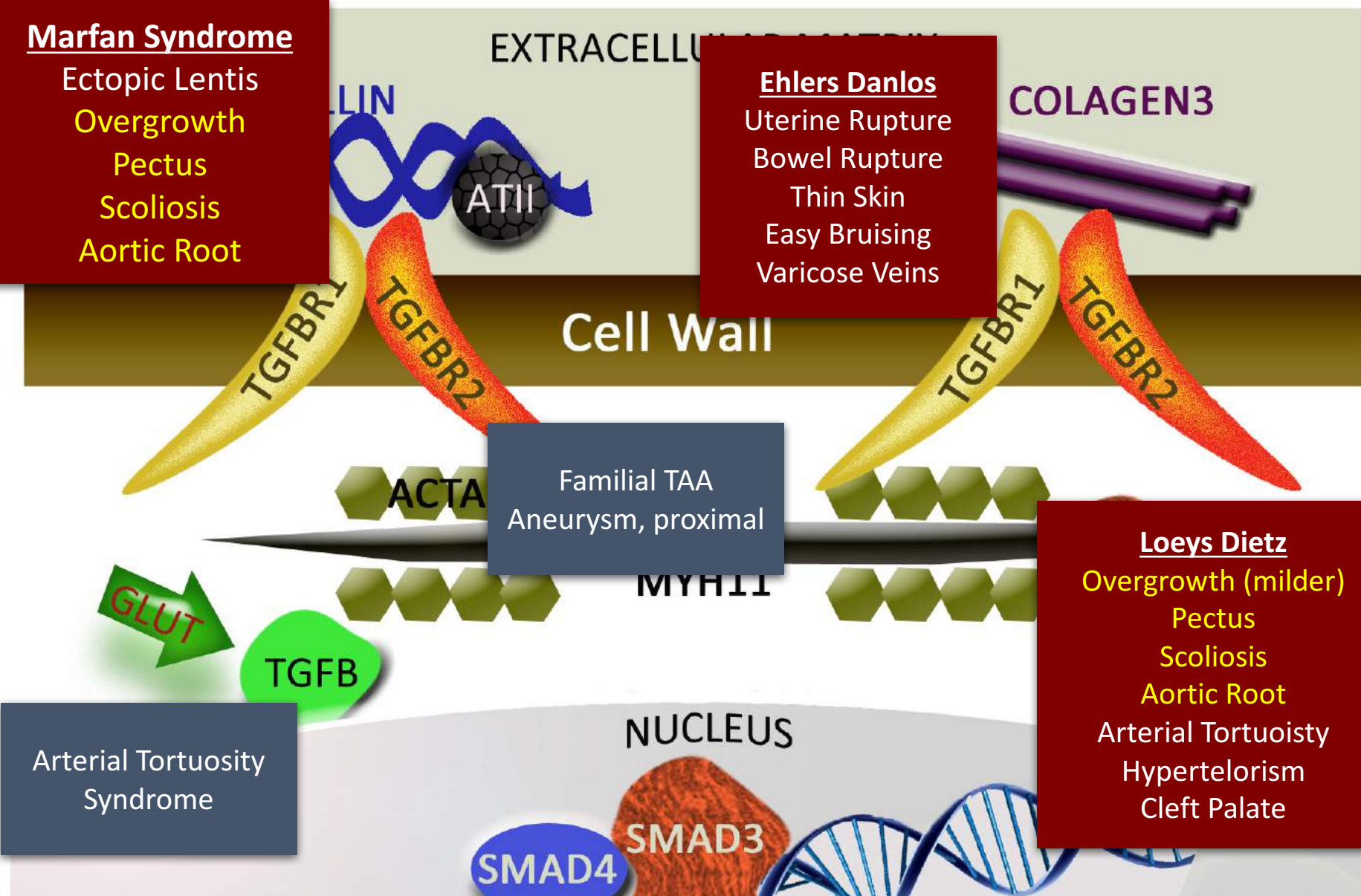
Uterine Rupture
Bowel Rupture
Thin Skin
Easy Bruising
Varicose Veins

Loeys Dietz

Overgrowth (milder)
Pectus
Scoliosis
Aortic Root
Arterial Tortuosity
Hypertelorism
Cleft Palate

Familial TAA
Aneurysm, proximal

Arterial Tortuosity
Syndrome



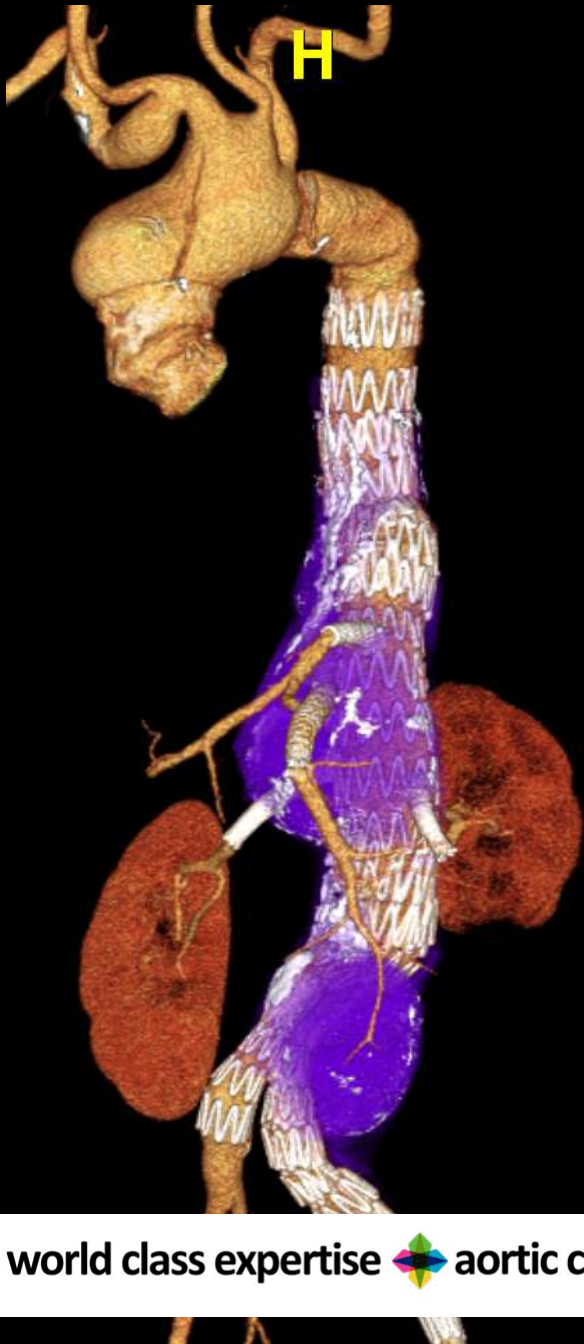


Age

Morphology

Family History





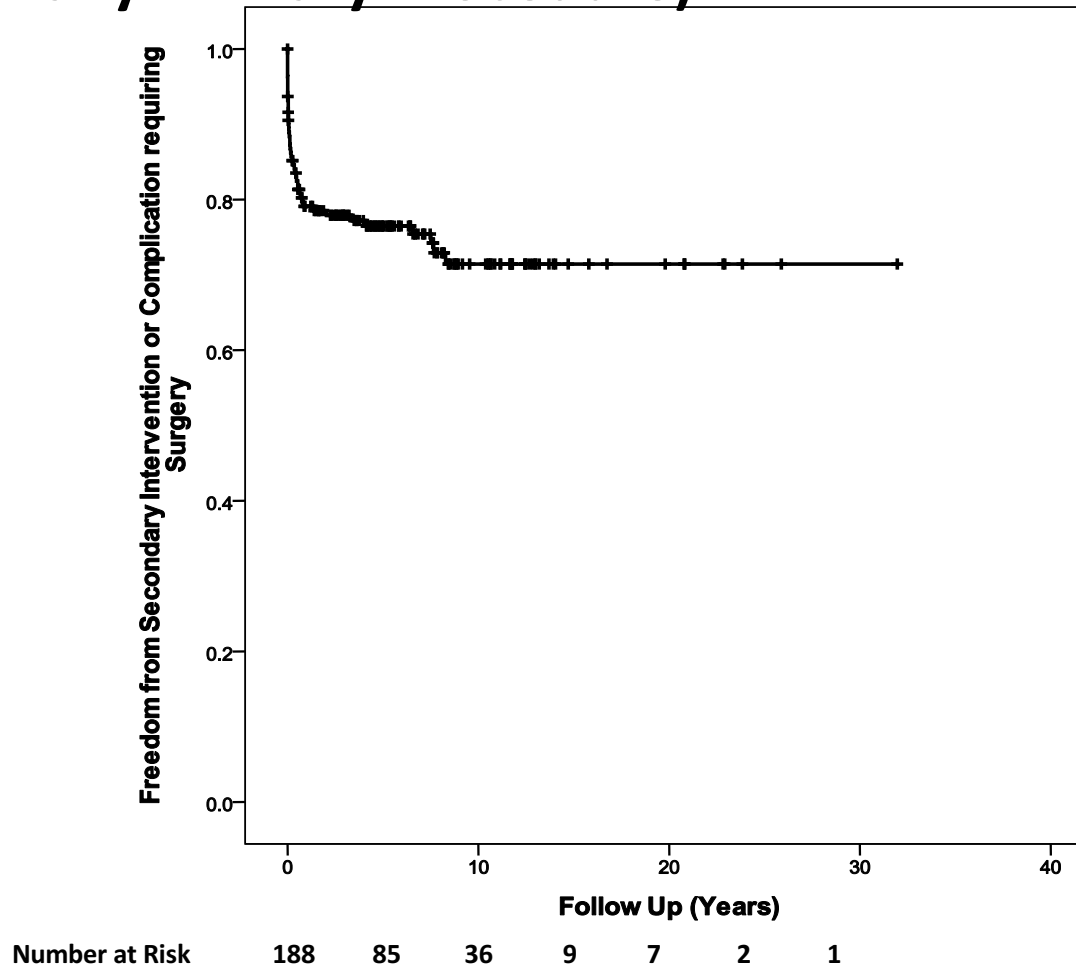
What do we know?

TEVAR and CTD

Author	Year	Pathology	N	Endoleak	Mortality (IH)	Stroke	SCI	Late intervention
Pacini	2013	Dissection	54	22%	2.50%	1.90%		
Eid-Lidt	2013	Dissection	10	44%	11%			33%
Waterman	2012	Dissection	16					
Preventza	2013	Dissection and Aneurysm	60		3.30%	1.6%	1.6%	17%

Freedom from Secondary Procedures

(After any Primary Procedure)



The Cleveland Experience

Beyond the Aortic Root: Staged Open and Endovascular Repair of Arch and Descending Aorta in Patients with Connective Tissue Disorders

Eric E. Roselli, MD,* Jay J. Idrees,* Ashley M. Lowry, MS, Khalil Masabni, MD, Edward G. Soltesz, MD, MPH, Douglas R. Johnston, MD, Vidyasagar Kalahasti, MD, Eugene H. Blackstone, MD, Joseph F. Sabik, III, MD, Bruce W. Lytle, MD, and Lars G. Svensson, MD, PhD

Aortic Center and Departments of Thoracic and Cardiovascular Surgery, and Cardiovascular Medicine Heart and Vascular Institute, Cleveland Clinic, Cleveland, Ohio; and Department of Quantitative Health Sciences, Research Institute, Cleveland Clinic, Cleveland, Ohio

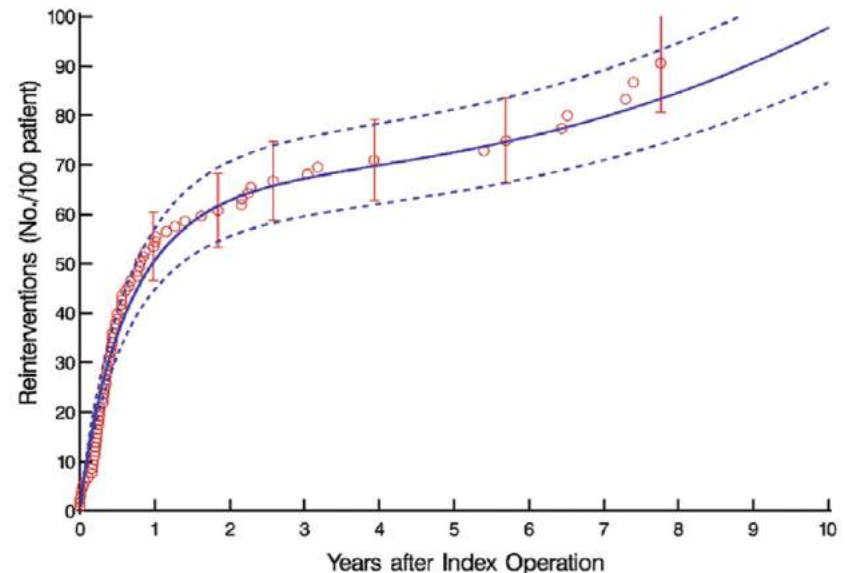
Background. Improvements in care have prolonged survival of patients with connective tissue disorders (CTDs), but their entire native aorta remains at risk. Little data are available to guide treatment. Objectives were to characterize patients, describe repair methods, and assess outcomes.

Methods. From 1996 to 2012, 527 patients with CTDs underwent cardiovascular operations. Beyond the root, arch and descending repair was performed in 121 patients (23%) for aneurysm (n = 17), acute complicated dissection (n = 5), or chronic dissection with aneurysmal degeneration (n = 99). CTD diagnoses included Marfan

aortic repair [TEVAR] 3). Median follow-up was 4.4 years.

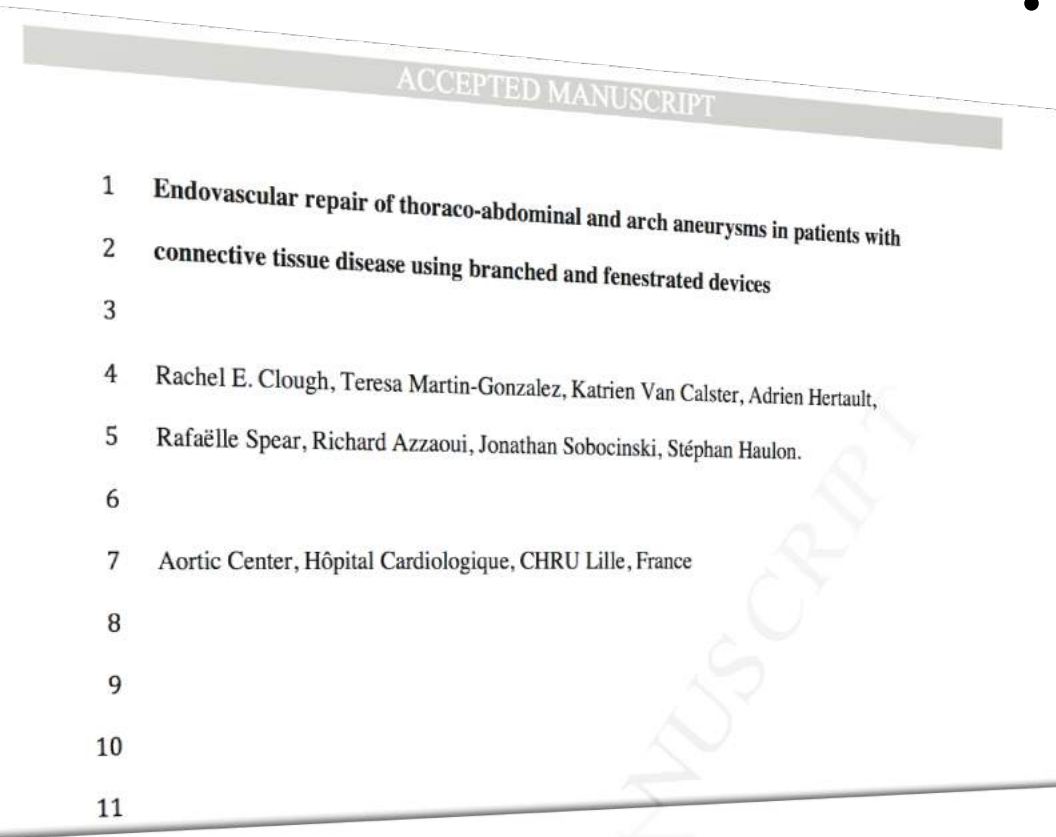
Results. Operative mortality was 2.5% (3 of 121). No paralysis occurred, but 3 patients (2.5%) had nonpermanent stroke, 4 (3.3%) required dialysis, 12 (10%) required tracheostomy, and 13 (11%) underwent reoperation for bleeding. During follow-up, 67 patients underwent 85 additional distal aortic procedures (58 open, 27 endovascular, 49 of which were stage II ET). By 10 years, probability of at least 1 reintervention was 61%. At 1, 5, and 10 years, estimated survival was 91%, 79%, and 62%, and event-free survival was 52%, 35%, and 24%, respectively.

- 1996 – 2012
- 527 Patients
- 1 Fenestrated Device



The Lille Experience

- 17/427 Patients
 - Mean age 51 years
 - Def'n CTD:
 - Met Marfan criteria
 - Family history
 - Extensive aortic disease at a young age
 - Joint hypermobility
 - Chest deformity
 - Characteristic facial features
 - Cleft palate
 - Bifid uvula
 - Hypertelorism
 - Translucent skin
- 100% technical success
- Follow up: 3.4 years
- 6/17: 35% aneurysm regression



Royal Free Experience: 2010-2017

	Age at Operation	CTD	Acuity	Mortality	SCI	Reintervention
1	60	Unknown	Elective	Lost to f/u		
2	57	Unknown	Elective	Lost to f/u		
3	60	Unknown	Elective	Lost to f/u		
4	58	Unknown	Elective	Lost to f/u		
5	58	Unknown	Elective	Alive	No	endoleak -- onyx
6	58	Unknown	Elective	Lost to f/u		
7	60	Unknown	Elective	Lost to f/u		
8	56	**Infection	Urgent	Unknown	Yes	
9	53	Family History	Elective	Alive	No	No
10	58	Unknown	Elective	Alive	No	No
11	39	Takayasu	Urgent	In Death	Yes	Post open repair
12	58	Unknown	Elective	Alive	Yes	Dissection, Staged
13	59	Family History	Elective	Alive	No	Dissection, Staged
14	44	Loeys Dietz	Elective	Alive	No	Iliac Extension

Royal Free Experience: 2014 - 2017

	Age at Operation	CTD	Acuity	Mortality	SCI	Reintervention
1	58	Unknown	Elective	Alive	No	endoleak -- onyx
2	56	**Infection	Urgent	Unknown	Yes	
3	53	Family History	Elective	Alive	No	No
4	58	Unknown	Elective	Alive	No	No
5	39	Takayasu	Urgent	In Hosp Death	Yes	Post open repair
6	58	Unknown	Elective	Alive	Yes	Dissection, Staged
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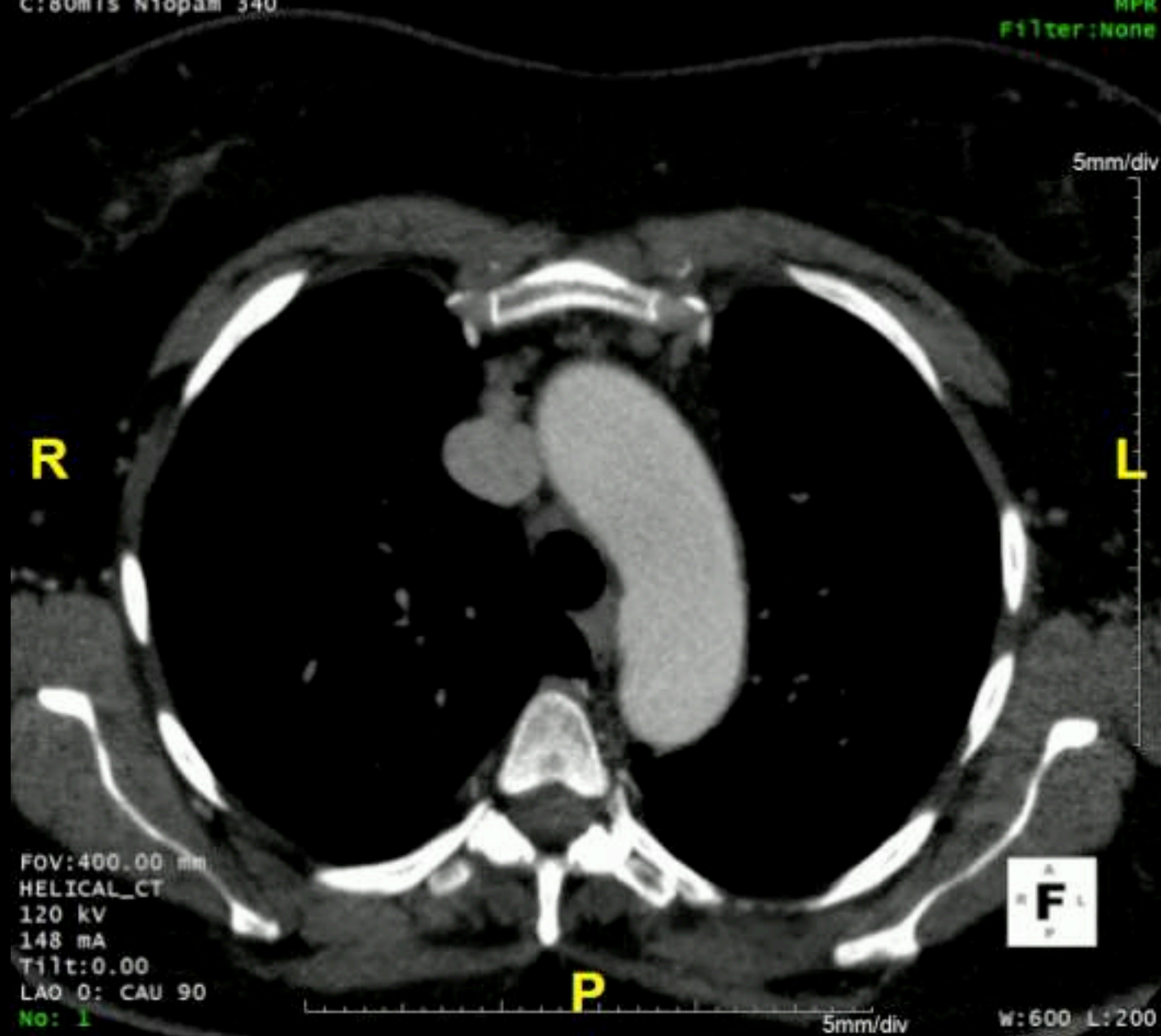
53 Year Old Woman

- No formal CTD diagnosis
- Healthy life to date
- Family History of Sudden Death
- Open attempt at repair, aborted at outside hospital
- ‘Too Scarred to proceed’
- Prolonged ITU stay, 6 months recovery
- Massive abdominal wall infection with skin grafting, now healed



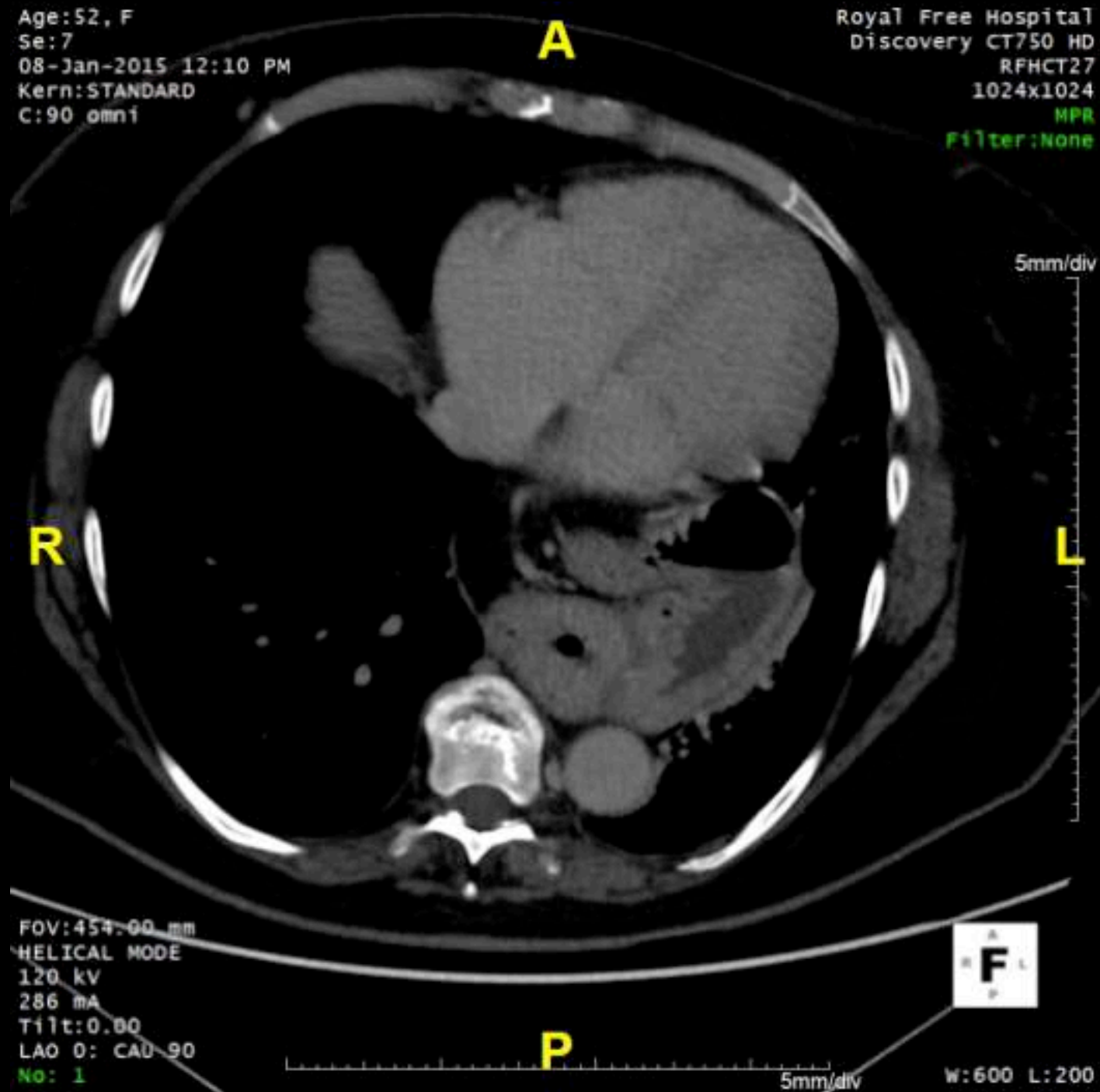
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Se: 4
18-Aug-2014 5:10 PM
Kern: FC13
C: 80mls Niopam 340

WORTHING HOSPITAL
Aquilion
burnbank
1024x1024
MPR
Filter: None



Age: 52, F
Se: 7
08-Jan-2015 12:10 PM
Kern: STANDARD
C: 90 omni

Royal Free Hospital
Discovery CT750 HD
RFHCT27
1024x1024
MPR
Filter: None



FOV: 454.00 mm
HELICAL MODE
120 kV
286 mA
Tilt: 0.00
LAO 0: CAU 90
No: 1

Age: 52, F
Se: 7
08-Jan-2015 12:10 PM
Kern: STANDARD
C: 90 omni

Royal Free Hospital
Discovery CT750 HD
RFHCT27
1024x1024
3D VR
Full

A

H

P

F

FOV: 454.00 mm
HELICAL MODE
120 kV
286 mA
Tilt: 0.00
LAO 90: CRA 0
No: 1

5mm/div



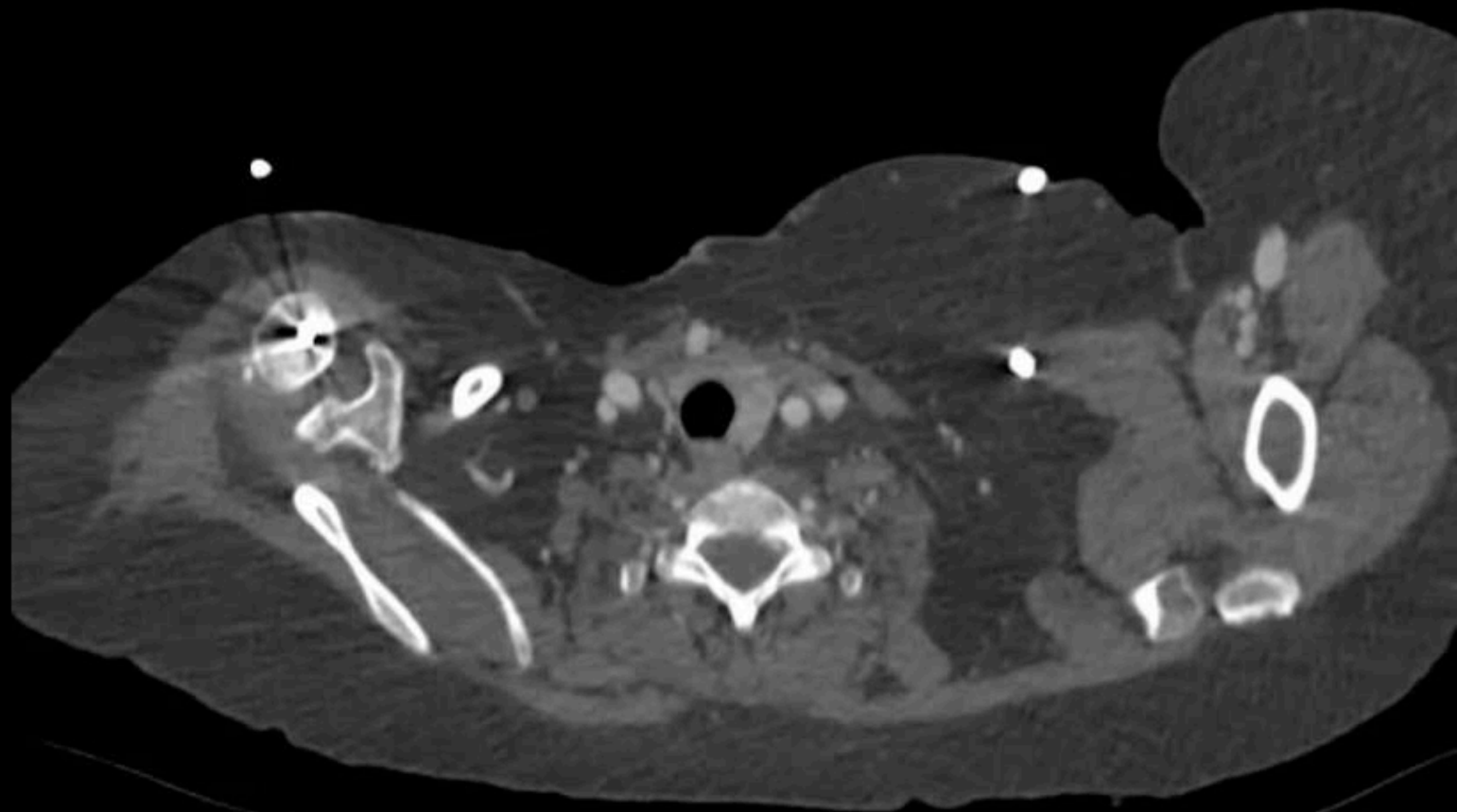
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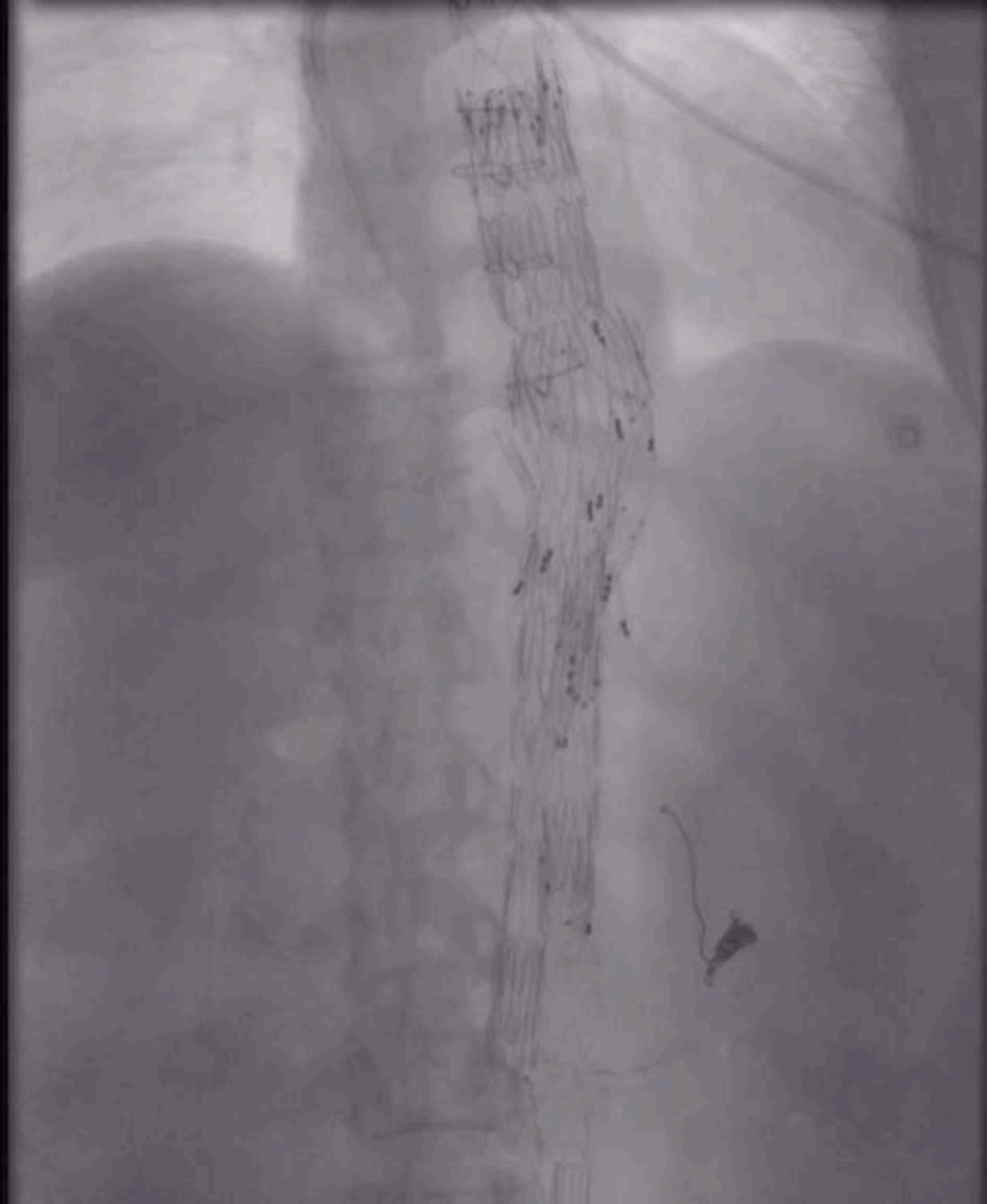


5mm/div

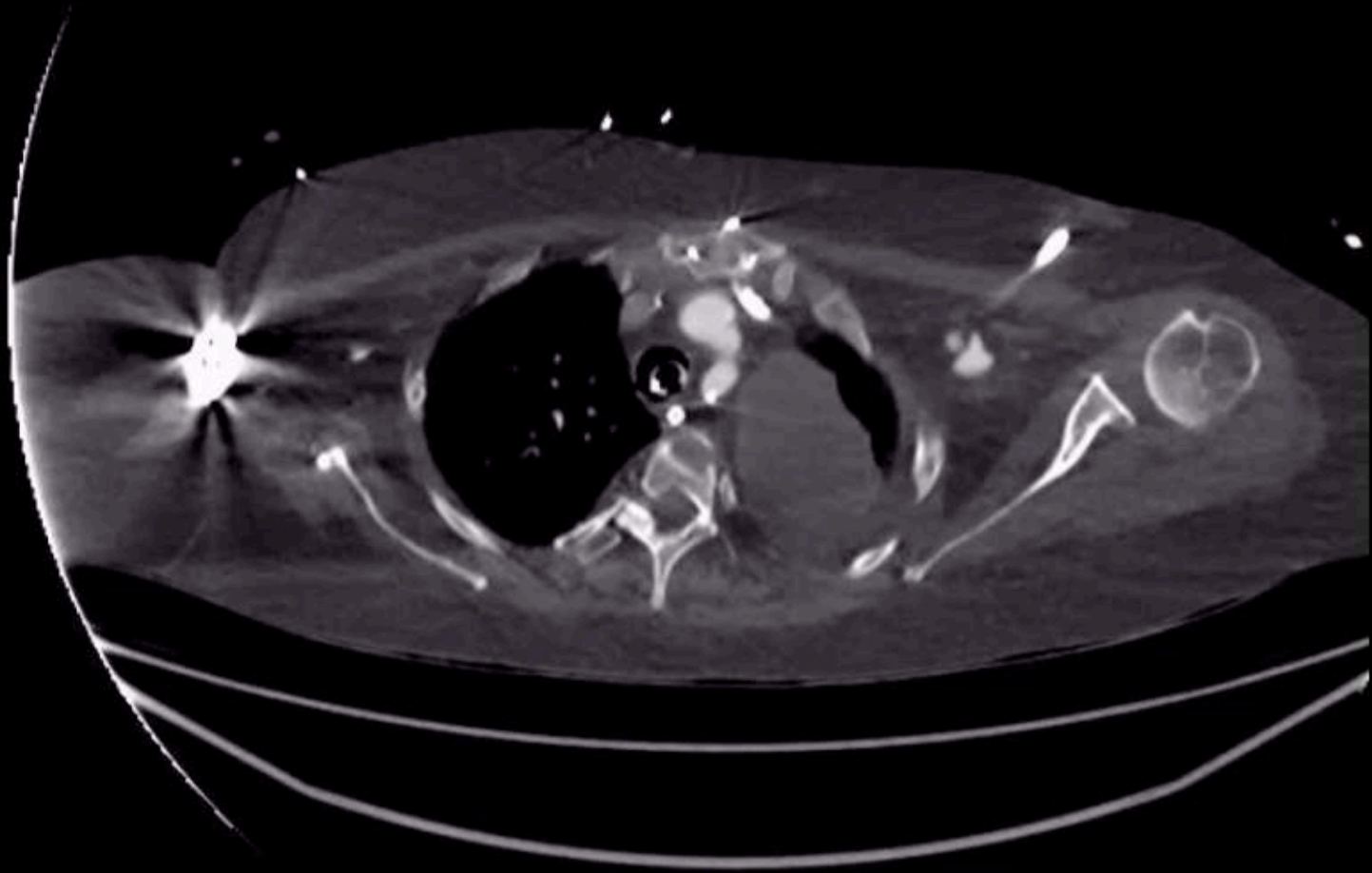
39 Year Old Woman

- “Takayasu” since young age
- Steroid dependant
- Morbidly Obese
- 20m Walking distance d/t SOB
- Ruptured AAA in Germany at age 32 – open repair
- Renal Failure on Dialysis
- **Presented with Symptomatic Aneurysm**





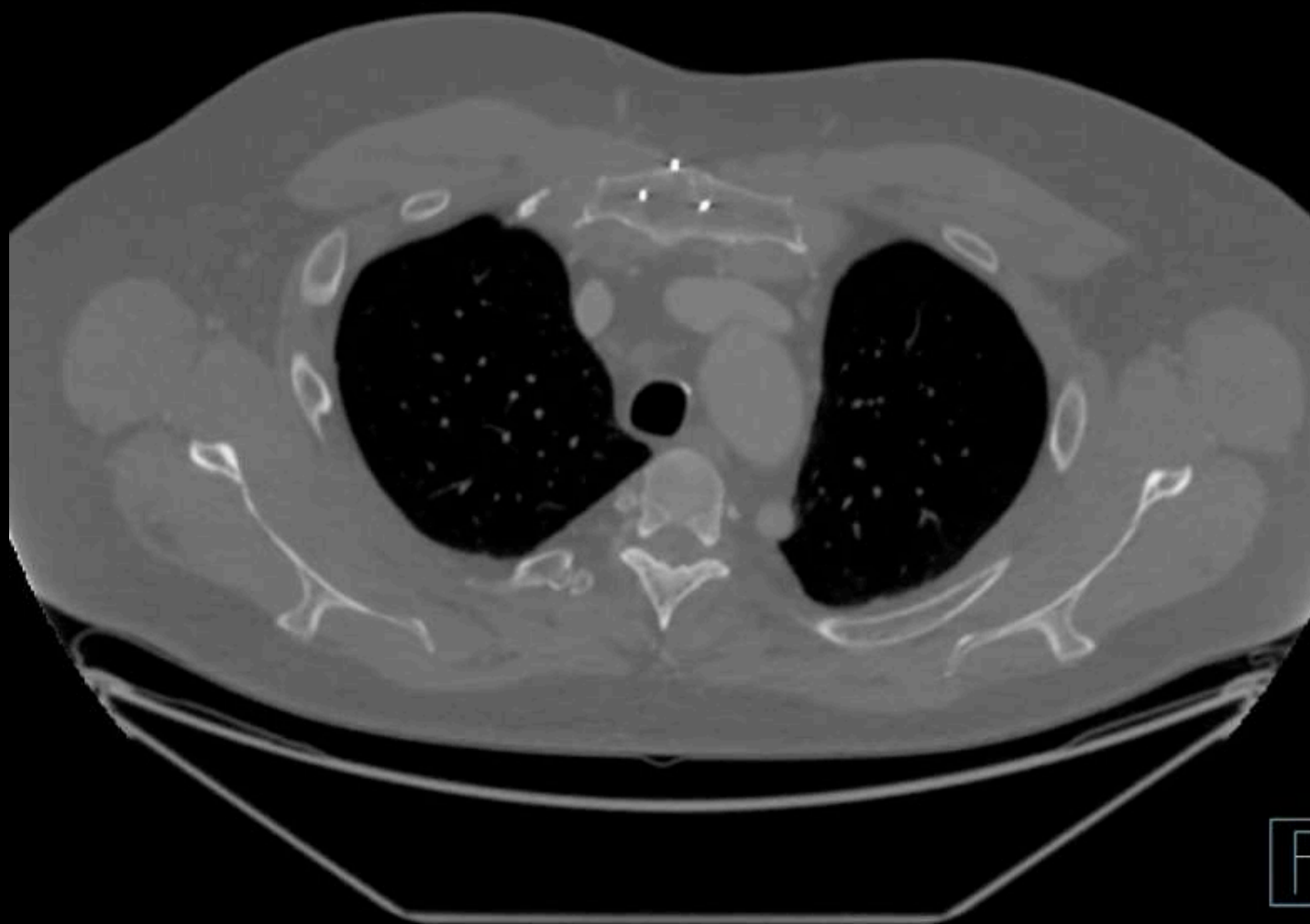
Spinal Cord Ischemia, Death at Day 20

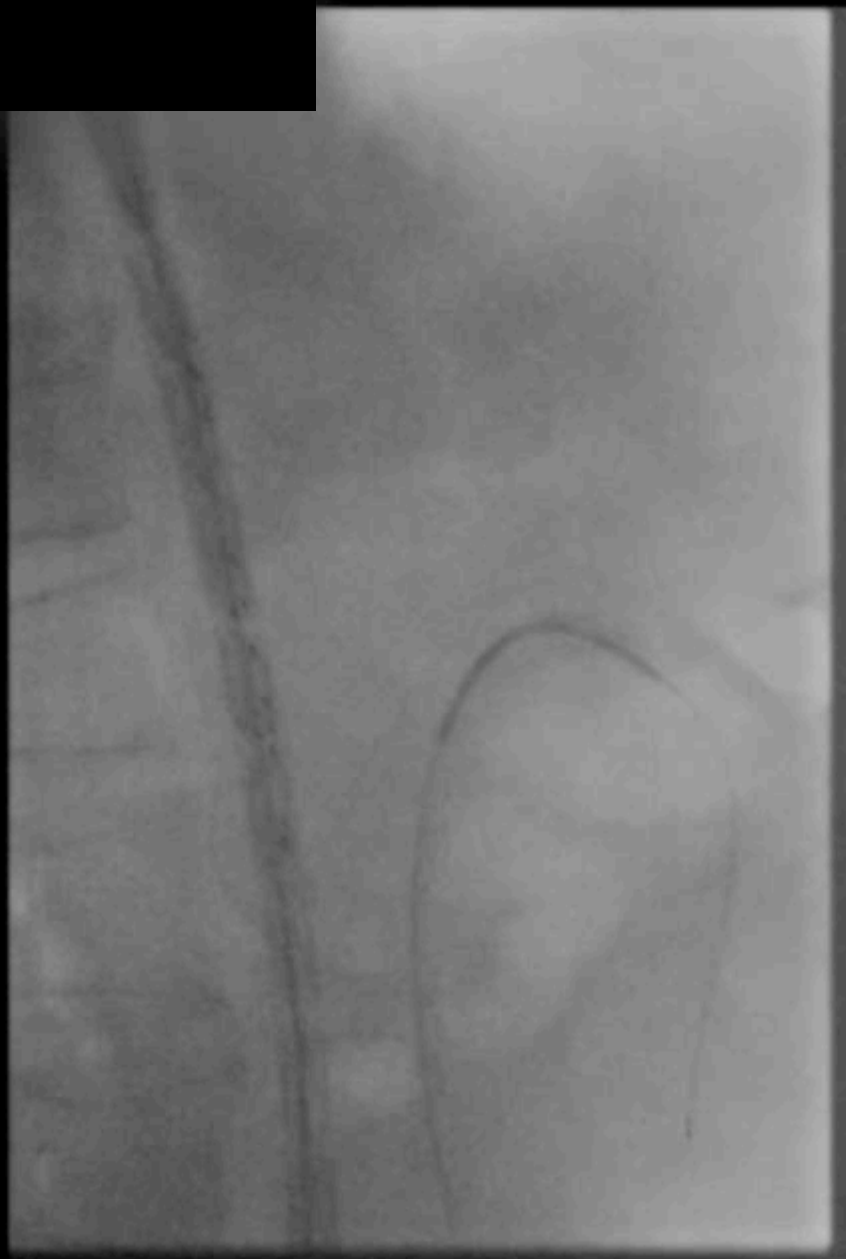


44 Year Old Man, 5.8cm JRAA

- Ureteric atresia requiring Mitrofanoff
- Ascending and St. Jude AVR 35 year old
- Renal transplant 36 years old
- Recurrent Urinary tract infections
- Morbid obesity

- Family history of fatal aortic rupture
- Dx Marfan/Loeys Dietz





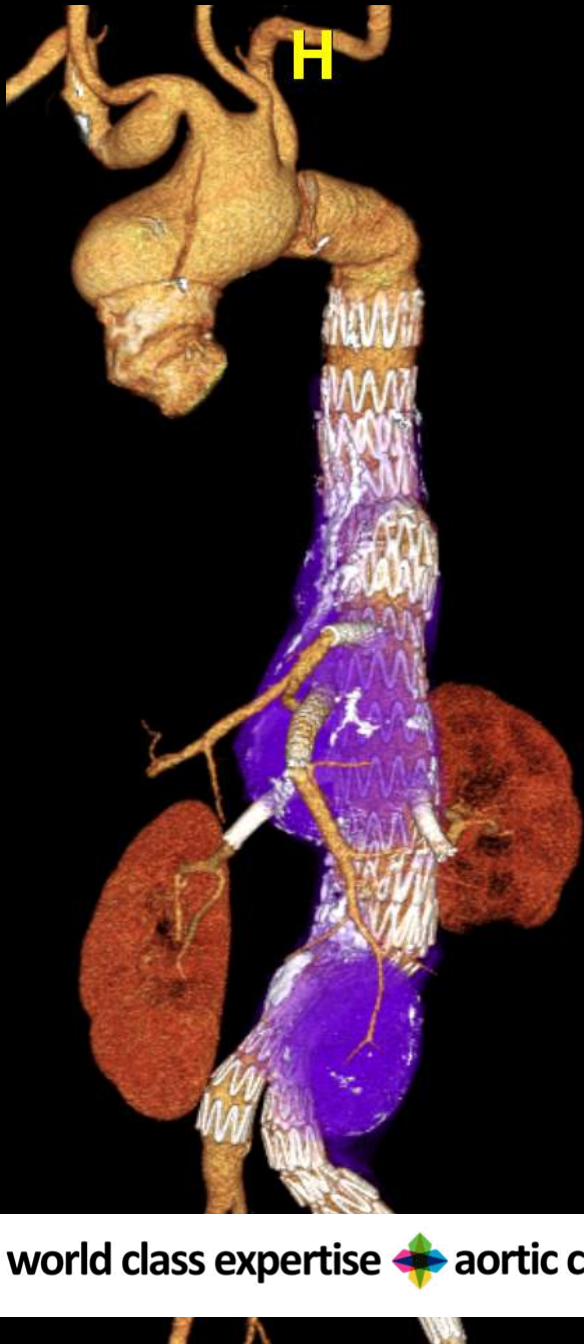
cm 42
RAO 63° / 0°

0030121
/2017
:06
A1



0
ot. 115

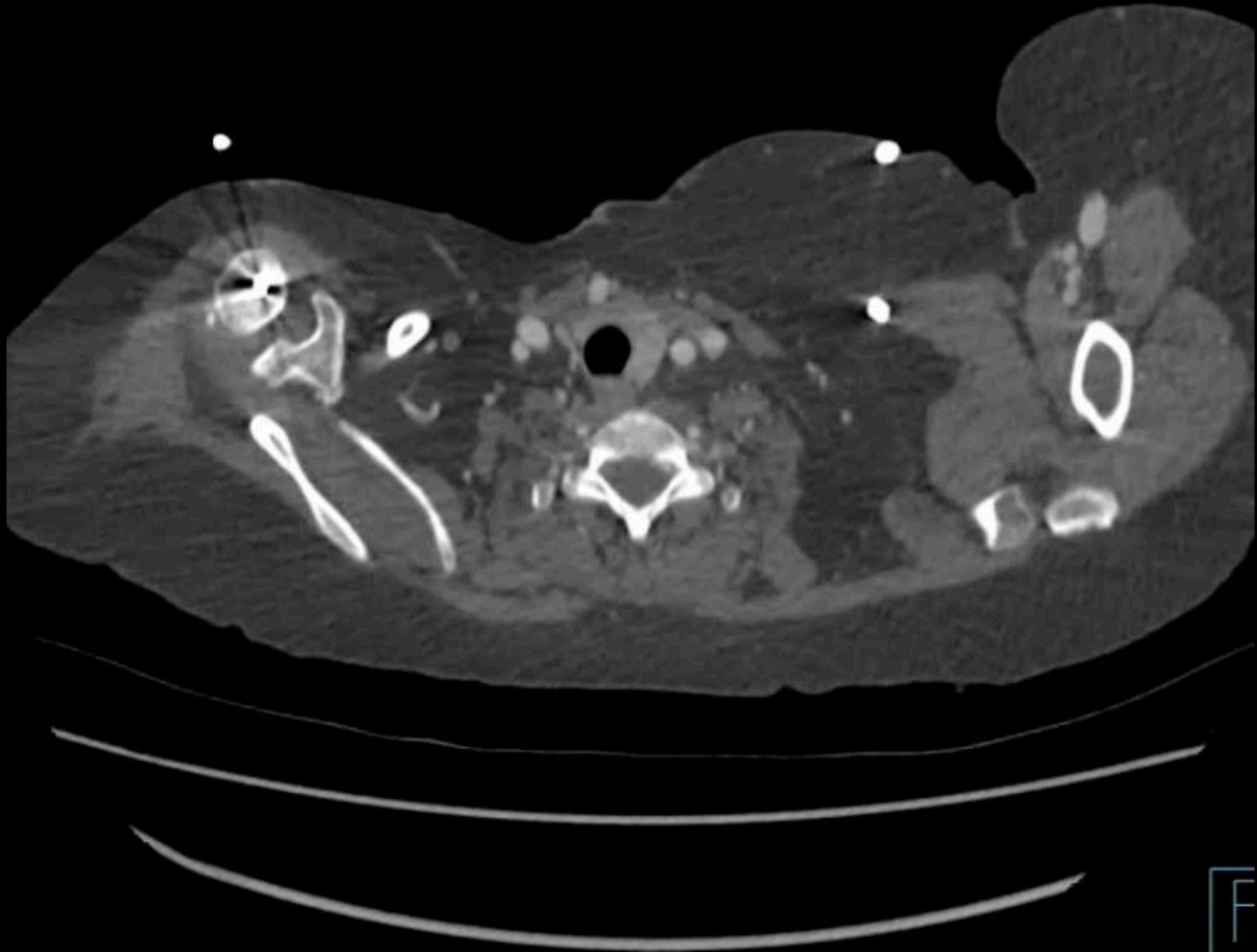
10
F
LCQ
W 10



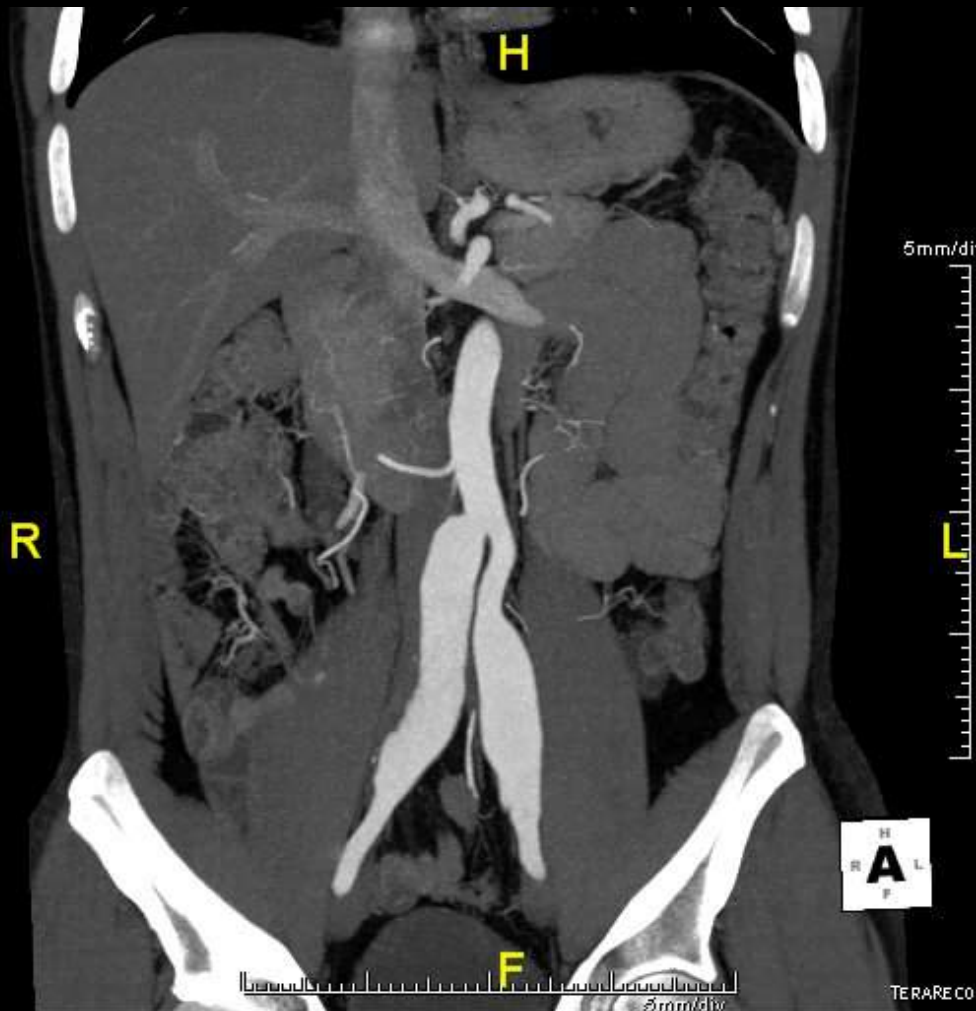
Are there Any Rules in CTD?

1. Trust No Artery
2. Access Problems are different
3. Not only aortic sealing zones fail
4. Talk to the Family
5. Rule out Infection
6. Be Creative

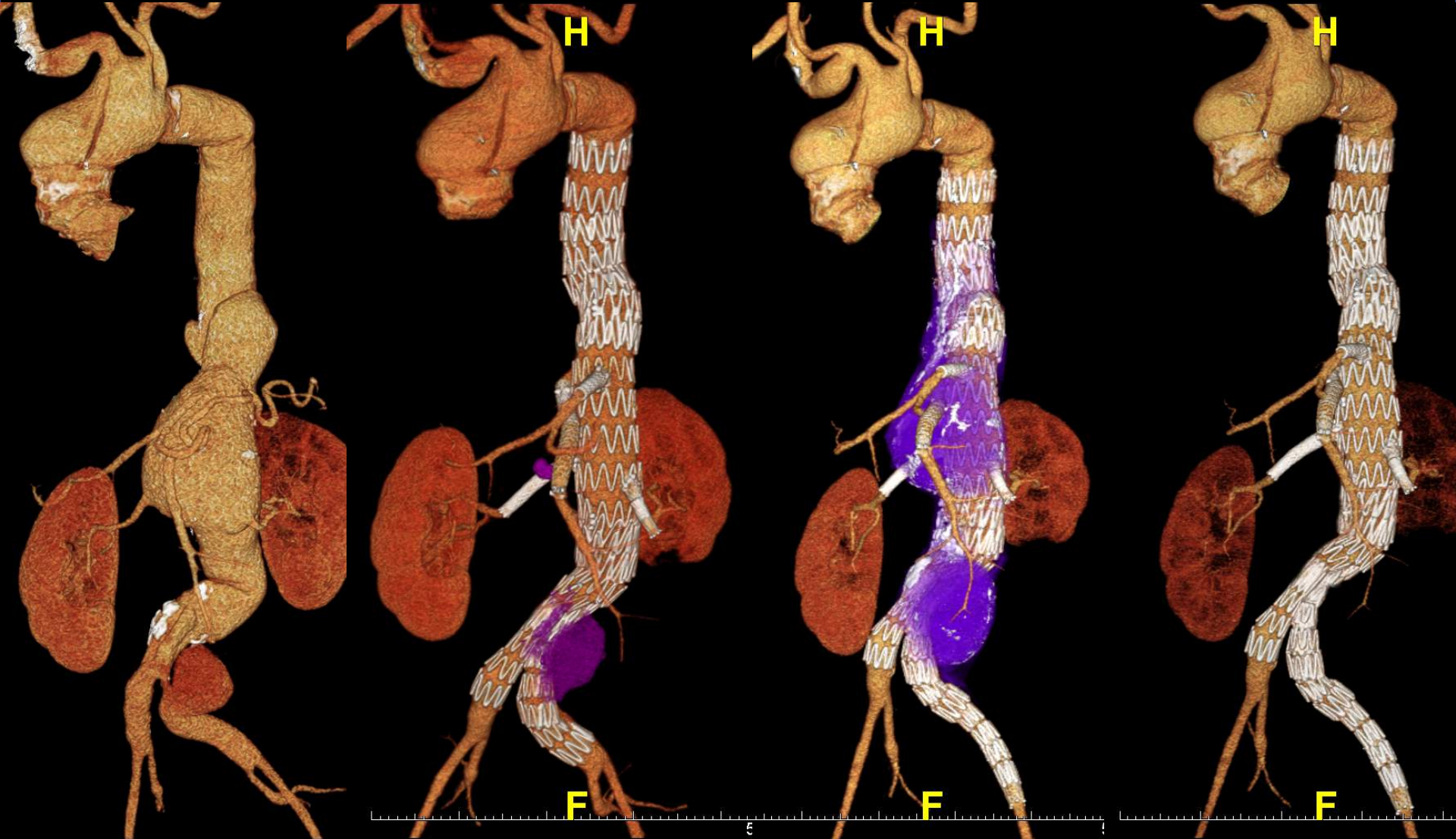
Trust No Artery...



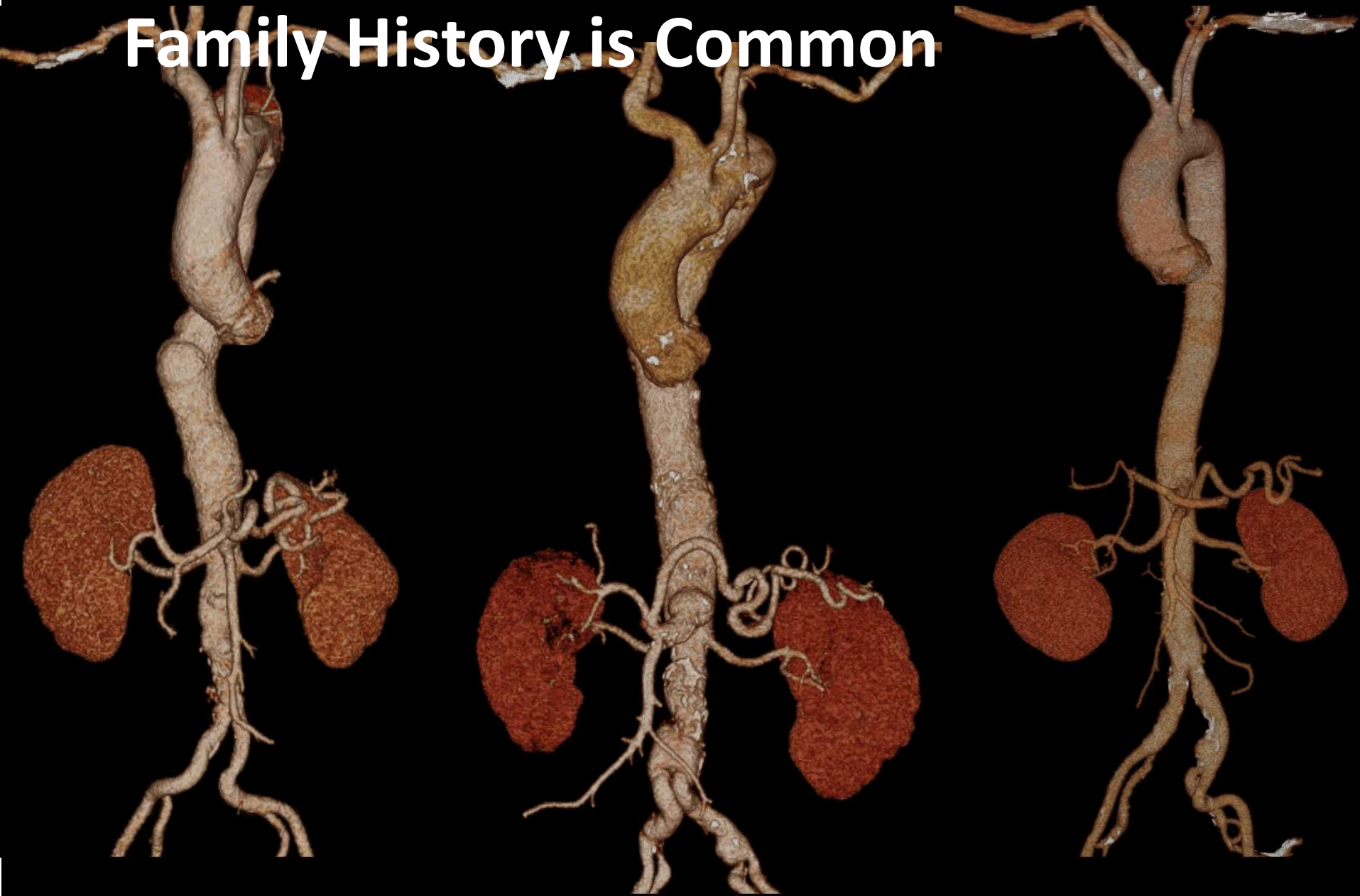
Access is Tricky



Aortic Sealing Zones are Not the Only Points of Failure

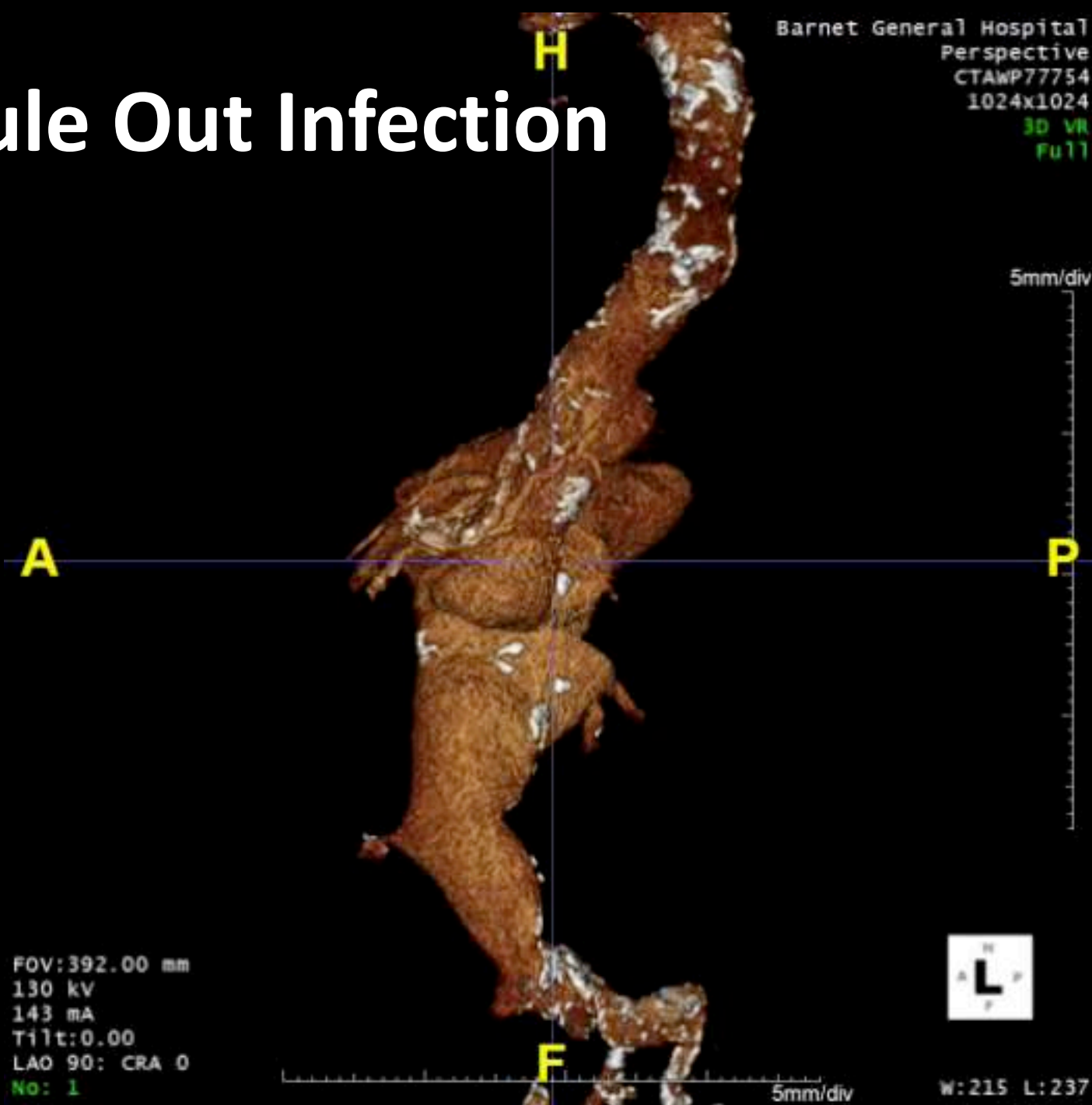


Family History is Common

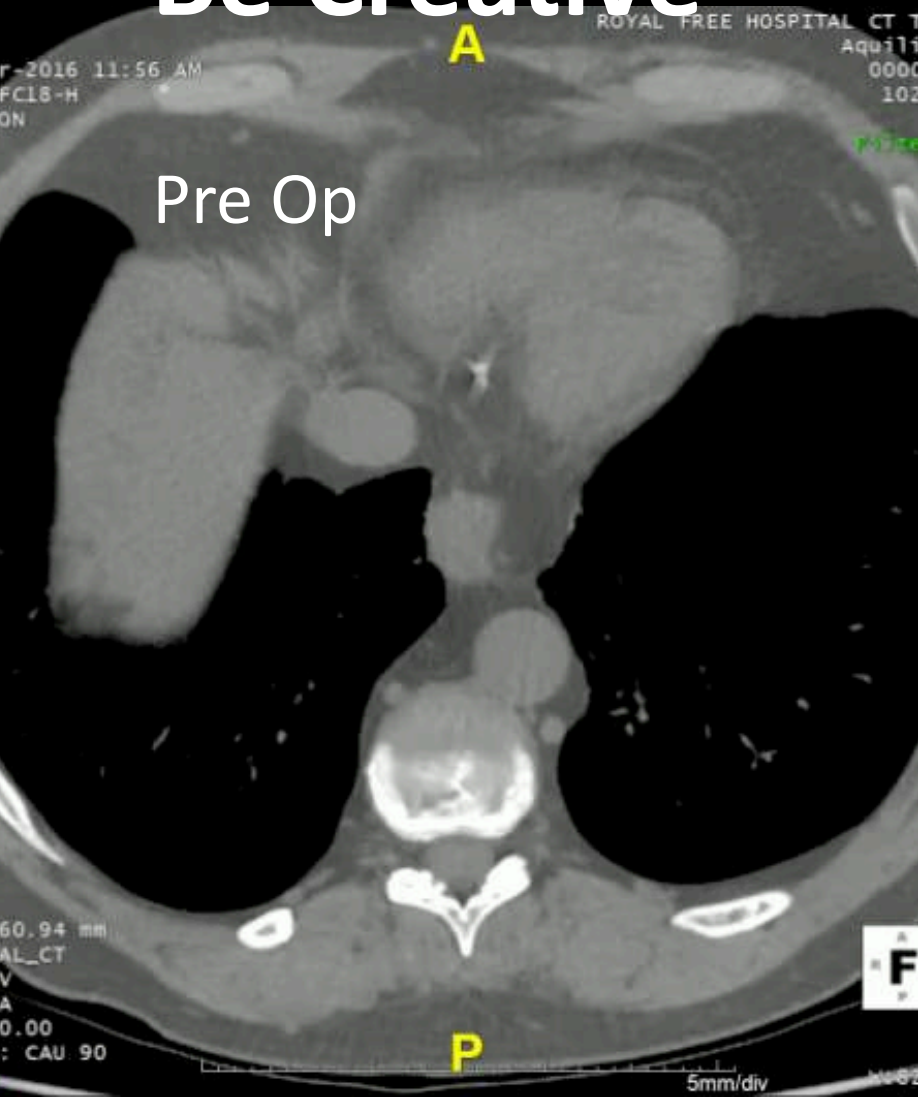


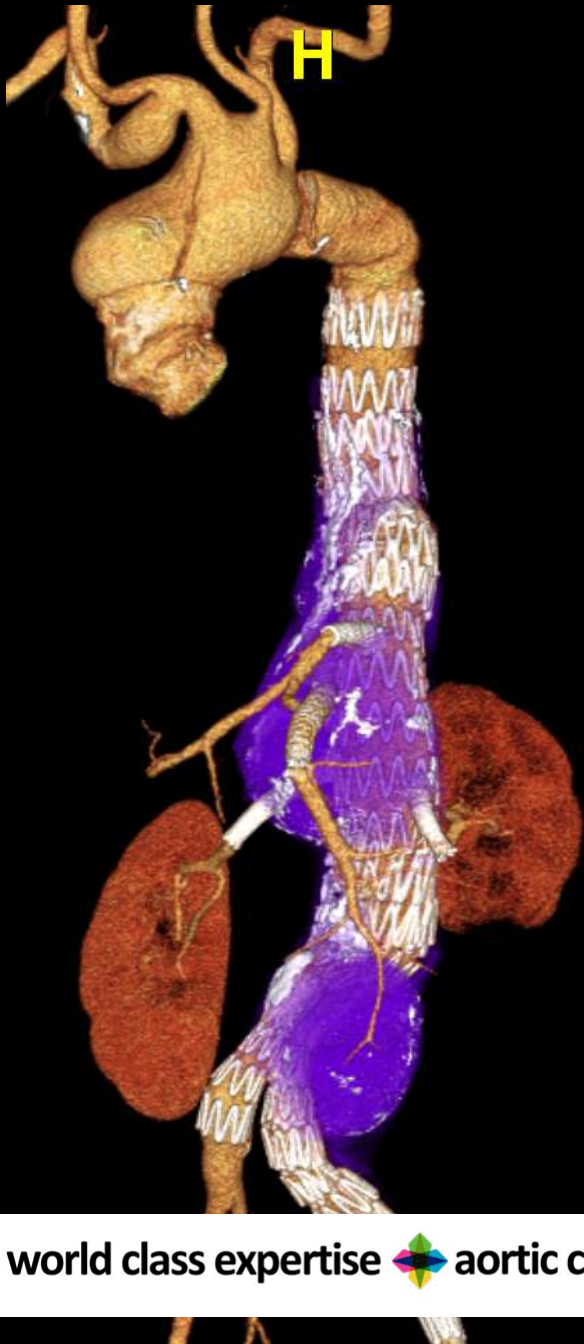
Rule Out Infection

Barnet General Hospital
Perspective
CTAWP77754
1024x1024
3D VR
Full



Be Creative





CTD And Complex Endovascular Repair

- Not standard of care
- Fraught with difficulty in the long and short term
- So much left to be understood

The Royal Free Aortic Team:

Meryl Davis

Debra Chong

Nick Schofield

Blandine Maurel

Teresa Martin Gonzalez

Prof George Hamilton

Myhrra Gazo

Yasmin Uddin

Vanessa Swann

Ved Romaini

world class expertise  aortic care