CKTCH
Centrum kardiovaskulární a transplantační chirurgie

Současné postavení PEARS (Exovascu) v léčbě výdutí kořene aorty

Petr Fila

CKTCH Brno

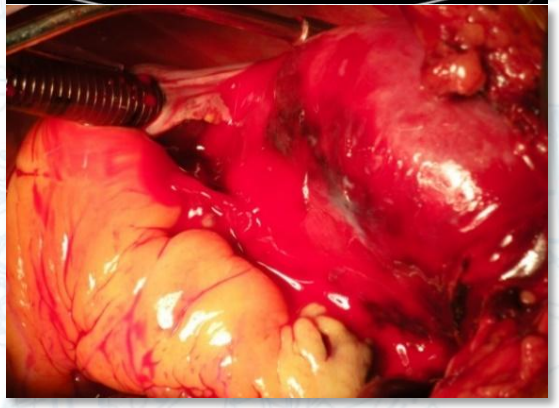
PEARS - myšlenka na individualiozované řešení

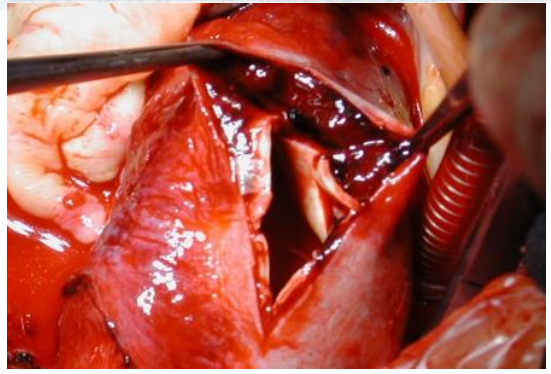
Tal Golesworthy - Marfanův syndrom

- PEARS - Personalised External Aortic Root Support
- polyesterová síťka kolem kořene a asc. aorty
- individualizace tvaru dle CT
- preventivní/kurativní výkon

Aneurysma aorty – co je cílem léčby?

Cíl? Zabránit dilataci/disekci! typu A - vzestupné aorty





- 20-50% zemře než se dostane do nemocnice/kardiochirurgii

Mahase, BMJ 2020; 368 :m304

- 50 % neléčených typ A zemře do 48 hod - mortalita 1-2 % / hod

- 30 denní mortalita po operaci 5-24%

Gudbjartsson,. Scandinavian Cardiovascular Journal, 2020, 54.1: 1-13.

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Doporučení

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GUIDELINES

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EACTS/STS Guidelines for diagnosing and treating acute and chronic syndromes of the aortic organ

Authors/Task Force Members: Martin Czerny [a,b,*,†] (Co-Chairperson) (Germany), Martin Grabenwöger[c,d,*,†] (Co-Chairperson) (Austria), Tim Berger[a,b] (Task Force Coordinator), Victor Aboyans[e,f] (France), Alessandro Della Corte [g,h] (Italy), Edward P. Chen[i] (USA), Nimesh D. Desai[j] (USA), Julia Dumfarth [k] (Austria), John A. Elefteriades[l] (USA), Christian D. Etz[m] (Germany), Karen M. Kim[n] (USA), Maximilian Kreibich[a,b] (Germany), Mario Lescan [o] (Germany), Luca Di Marco[p] (Italy), Andreas Martens [q,r] (Germany), Carlos A. Mestres [s] (South Africa), Milan Milojevic [t] (Serbia), Christoph A. Nienaber [u,v] (UK), Gabriele Piffaretti[w] (Italy), Ourania Preventza[x] (USA), Eduard Quintana[y] (Spain), Bartosz Rylski [a,b] (Germany), Christopher L. Schlett[b,z] (Germany), Florian Schoenhoff[aa] (Switzerland), Santi Trimarchi[ab] (Italy) and Konstantinos Tsagakis [ac] (Germany), EACTS/STS Scientific Document Group

ESC European Society of Cardiology

European Heart Journal (2024) **00**, 1–163
https://doi.org/10.1093/eurheartj/ehae179

ESC GUIDELINES

2024 ESC Guidelines for the management of peripheral arterial and aortic diseases

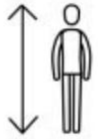
Developed by the task force on the management of peripheral arterial and aortic diseases of the European Society of Cardiology (ESC)

Authors/Task Force Members: Lucia Mazzolai [*†], (Chairperson) (Switzerland), Gisela Teixido-Tura [‡], (Task Force Co-ordinator) (Spain), Stefano Lanzi [‡], (Task Force Co-ordinator) (Switzerland), Vinko Boc (Slovenia), Eduardo Bossone (Italy), Marianne Brodmann [1] (Austria), Alessandra Bura-Rivière (France), Julie De Backer [2] (Belgium), Sebastien Deglise (Switzerland), Alessandro Della Corte (Italy), Christian Heiss (United Kingdom), Marta Kałużna-Oleksy (Poland), Donata Kurpas (Poland), Carmel M. McEniery (United Kingdom), Tristan Mirault (France), Agnes A. Pasquet (Belgium), Alex Pitcher (United Kingdom), Hannah A.I. Schaubroeck (Belgium), Oliver Schlager (Austria), Per Anton Sirnes (Norway), Muriel G. Sprynger (Belgium), Eugenio Stabile (Italy), Françoise Steinbach (France), Matthias Thielmann (Germany), Roland R.J. van Kimmenade (Netherlands), Maarit Venermo (Finland), Jose F. Rodriguez-Palomares [*†], (Chairperson) (Spain), and ESC Scientific Document Group

***Risk factors (RF)**

Length of ascending aorta ≥11cm

>3mm diameter increase per year



Height <1.69m

Age <50 years old

Arterial hypertension

Czerny, European Journal of Cardio-Thoracic Surgery, 2024, 65.2: ezad426.
Mazzolai, European heart journal, 2024, 45.36: 3538-3700.

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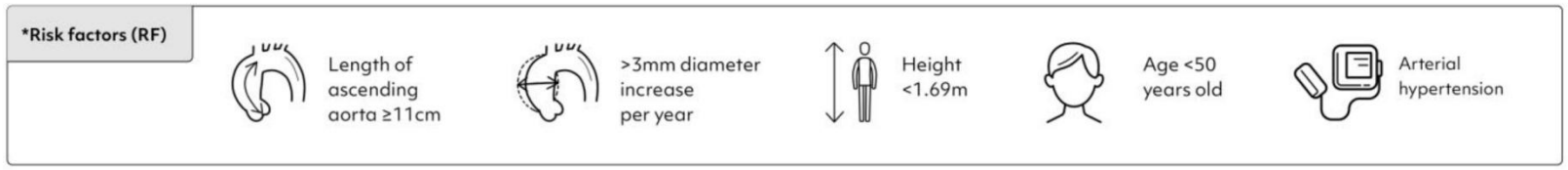
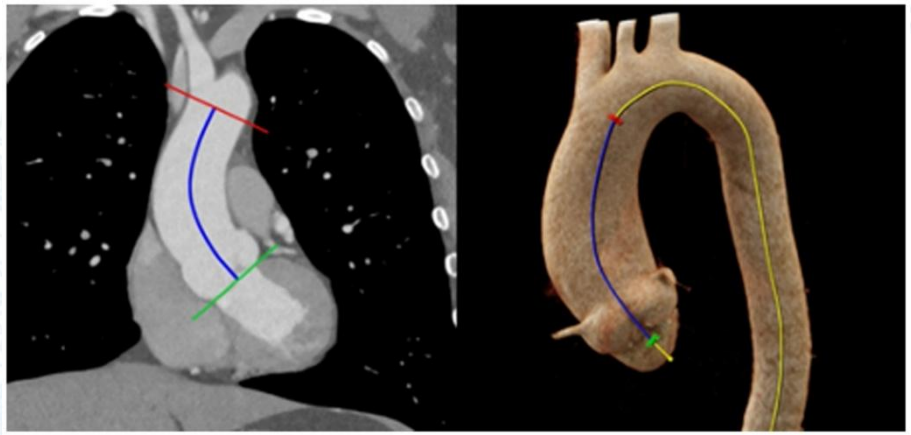
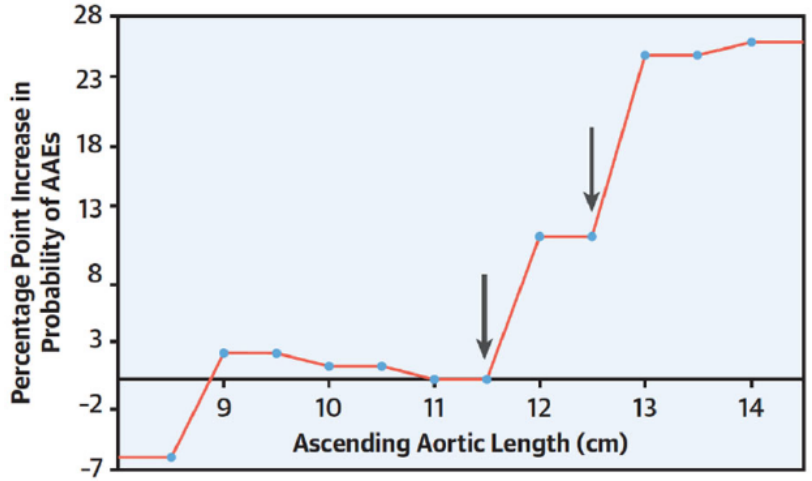
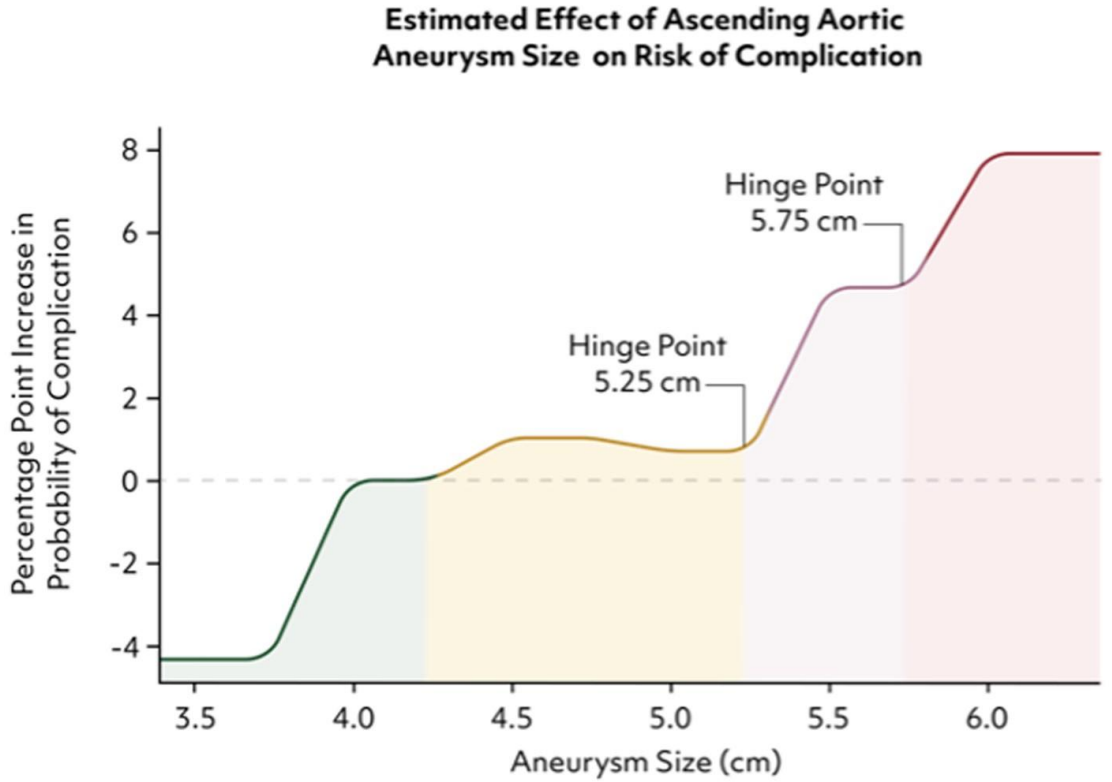
Chirurgická léčba









Czerny, European Journal of Cardio-Thoracic Surgery, 2024, 65.2: ezad426.
Della Corte A, Eur J Cardiothorac Surg 2021;60:978–86.

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Rizikové faktory disekce

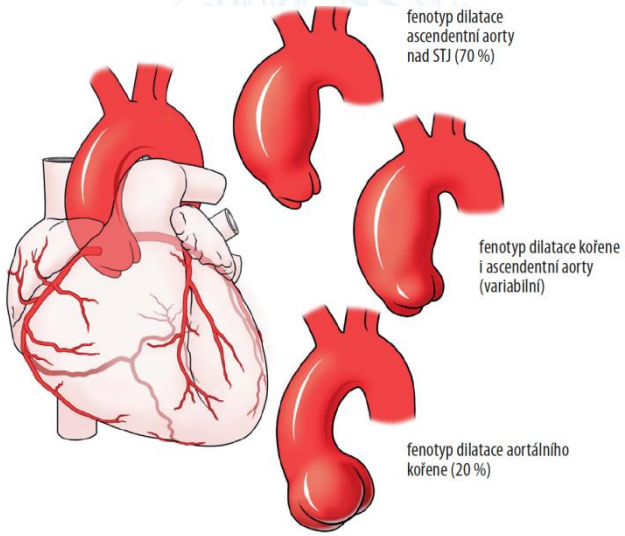
- hypertenze
- bikuspidní Ao chlopeň
- degenerace/zánět onemocnění aortální stěny
- ateroskleróza
- **geneticky podmínění aortopatie**
  Marfan, Ehlers-Danlos, Loeys-Dietz syndrom, Turner ... ACTA2, MYH11, MYLK, PRKG…
- tupé trauma
- polycystická choroba ledvin
- koarktace
- iatrogenní
- …

spojené s vysokou vaskulární morbiditou/mortalitou
mladí
bez přítomnosti klasických RF

indikace k operačnímu řešení:
agresivita mutace
dynamika růstu aorty
RA disekce v mladém věku
fenotyp aorty

Nejen dilatace/rozměr aorty!

Hranice pro výkon na aortě



- Marfan + RF (IIa)
- TGFBR 1,2 mutace
- konkomitantní výkon

- Marfan (I)
- BAV root (I)
- BAV asc. + RF (IIa)
- TAV low-risk(IIa)

- izolované aneurysma
- BAV asc. (I)
- TAV root (I)

40 mm **42 mm** **45 mm** **50 mm** **52 mm** **55 mm**

- PRKG1 + RF

- ACT2 + RF
- PRKG1

- ACTA2
- MYLK, MYH11

- TAV asc. (IIa)

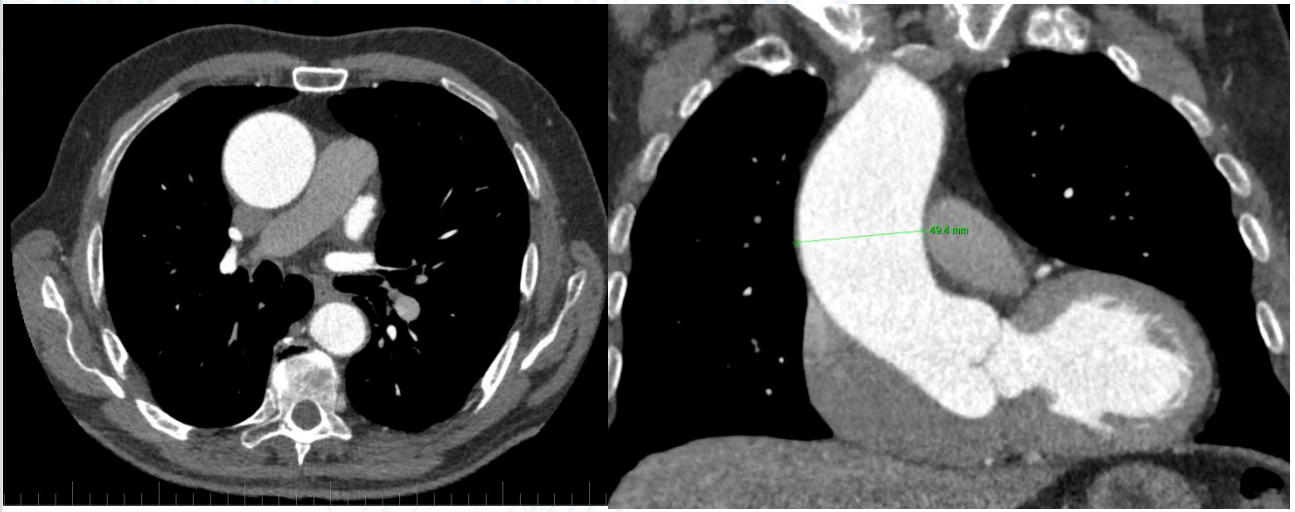
- Loeys-Dietz syndrom (40-50mm) – podle patogenní varianty

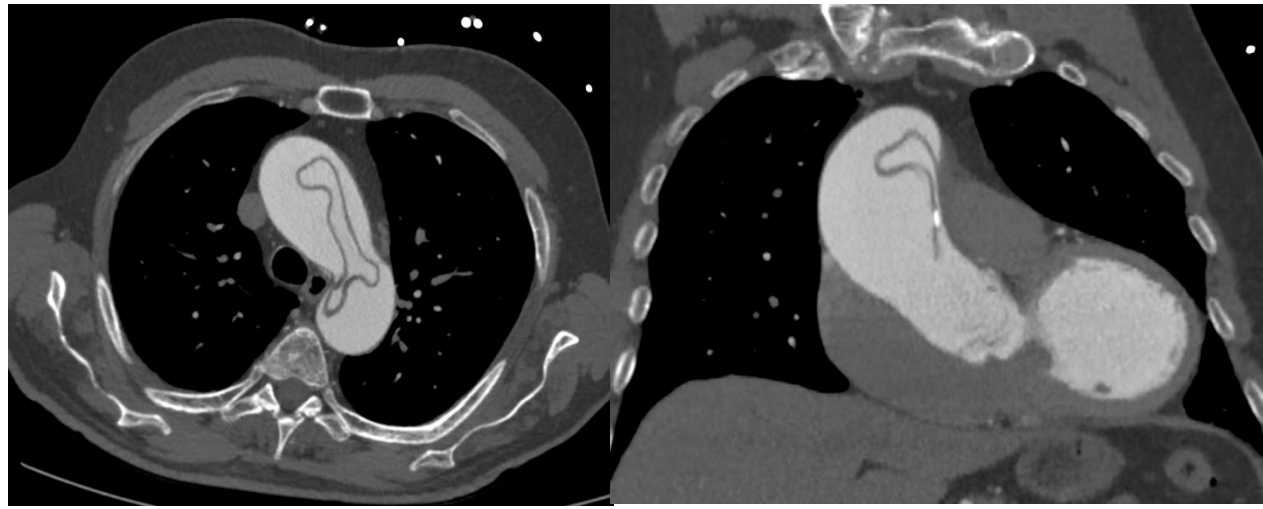
Hranice pro výkon na aortě

40 mm	42 mm	45 mm	50 mm	52 mm	55 mm
▪ PRKG1 + RF	▪ ACT2 + RF ▪ PRKG1	▪ Marfan + RF (IIa) ▪ TGFBR 1,2 mutace ▪ konkomitantní výkon	▪ Marfan (I) ▪ BAV root (I) ▪ BAV asc. + RF (IIa) ▪ TAV low-risk(IIa)	▪ TAV asc. (IIa)	▪ izolované aneurysma ▪ BAV asc. (I) ▪ TAV root (I)
		▪ ACTA2 ▪ MYLK, MYH11			

▪ Loeys-Dietz syndrom (40-50mm) – podle patogenní varianty

… o rok později…





Predikce disekce aorty typu A

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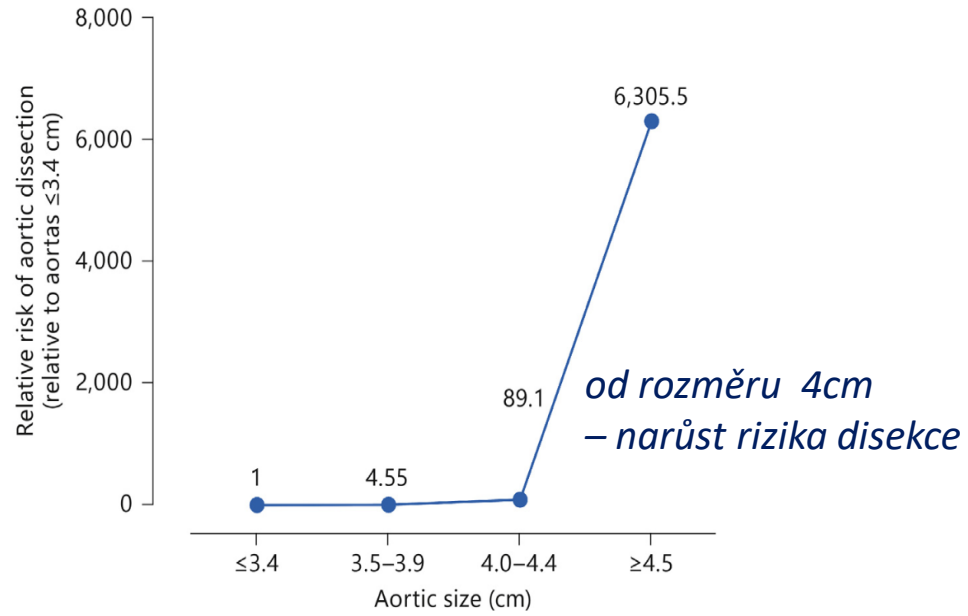
CLINICAL PRACTICE GUIDELINE

2022 ACC/AHA Guideline for the Diagnosis and Management of Aortic Disease

FIGURE 5 Relative Risk of Aortic Dissection by Size Range



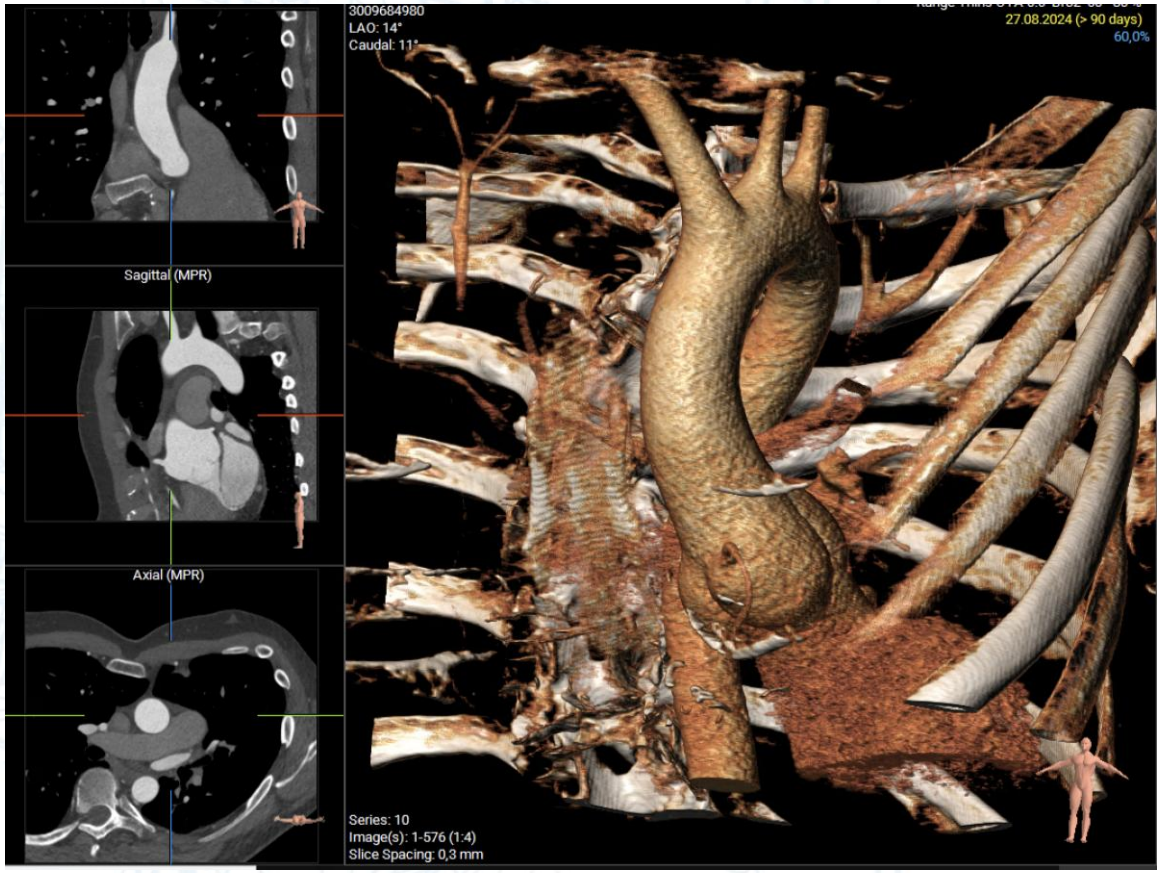
od rozměru 4cm – narůst rizika disekce

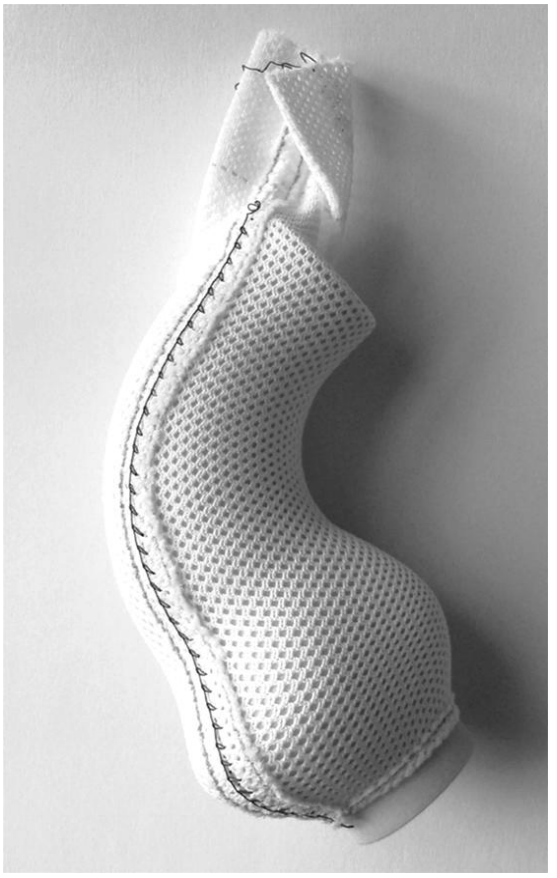
The relative risk of aortic dissection begins to increase appreciably at a diameter of 4.0 cm to 4.4 cm and then increases dramatically at a diameter of ≥4.5 cm. Reprinted from Paruchuri et al.[5] Copyright 2005, with permission from Karger Publishers, Basel Switzerland.

- In asymptomatic patients with aneurysms of the aortic root or ascending aorta who have a maximum diameter of ≥5.0 cm, surgery is reasonable when performed by experienced surgeons in a Multidisciplinary Aortic Team.[14-17]

Isselbacher, JACC, 2022, 80.24: e223-e393.
Paruchuri, Cardiology, 2015, 131.4: 265-272.

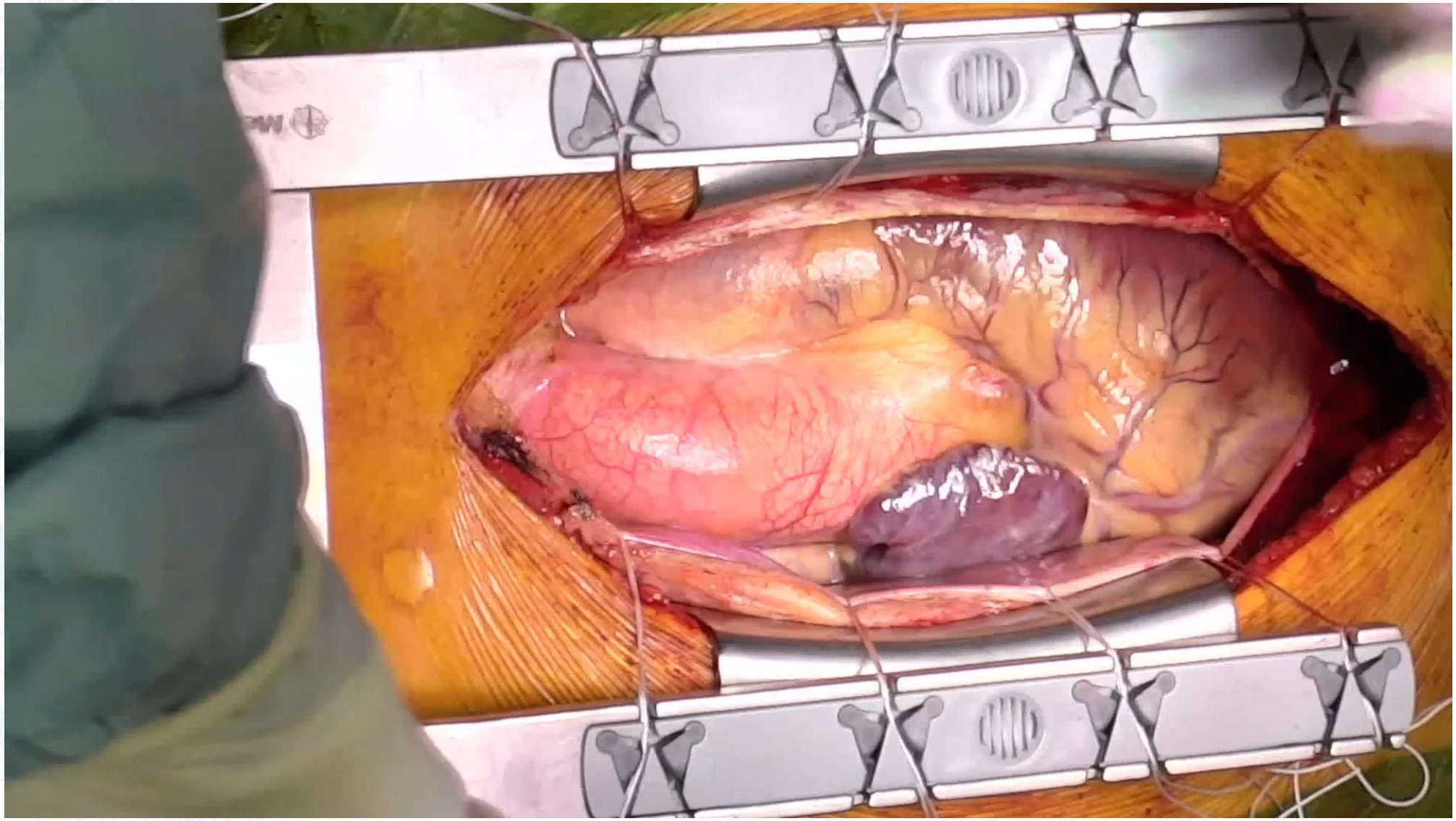
Posun k preventivní chirurgické léčbě - individualizované řešení



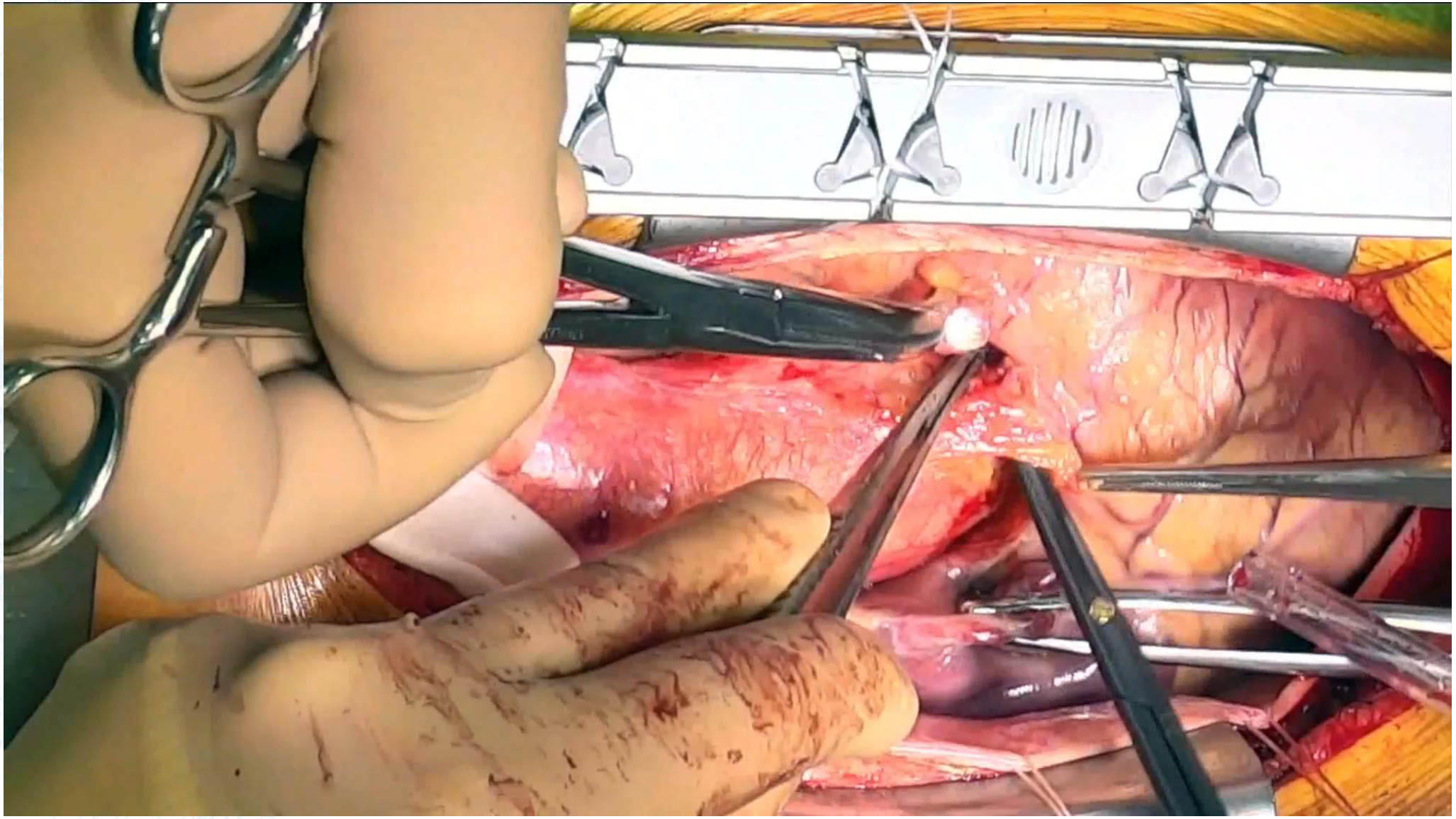


Individualizované řešení

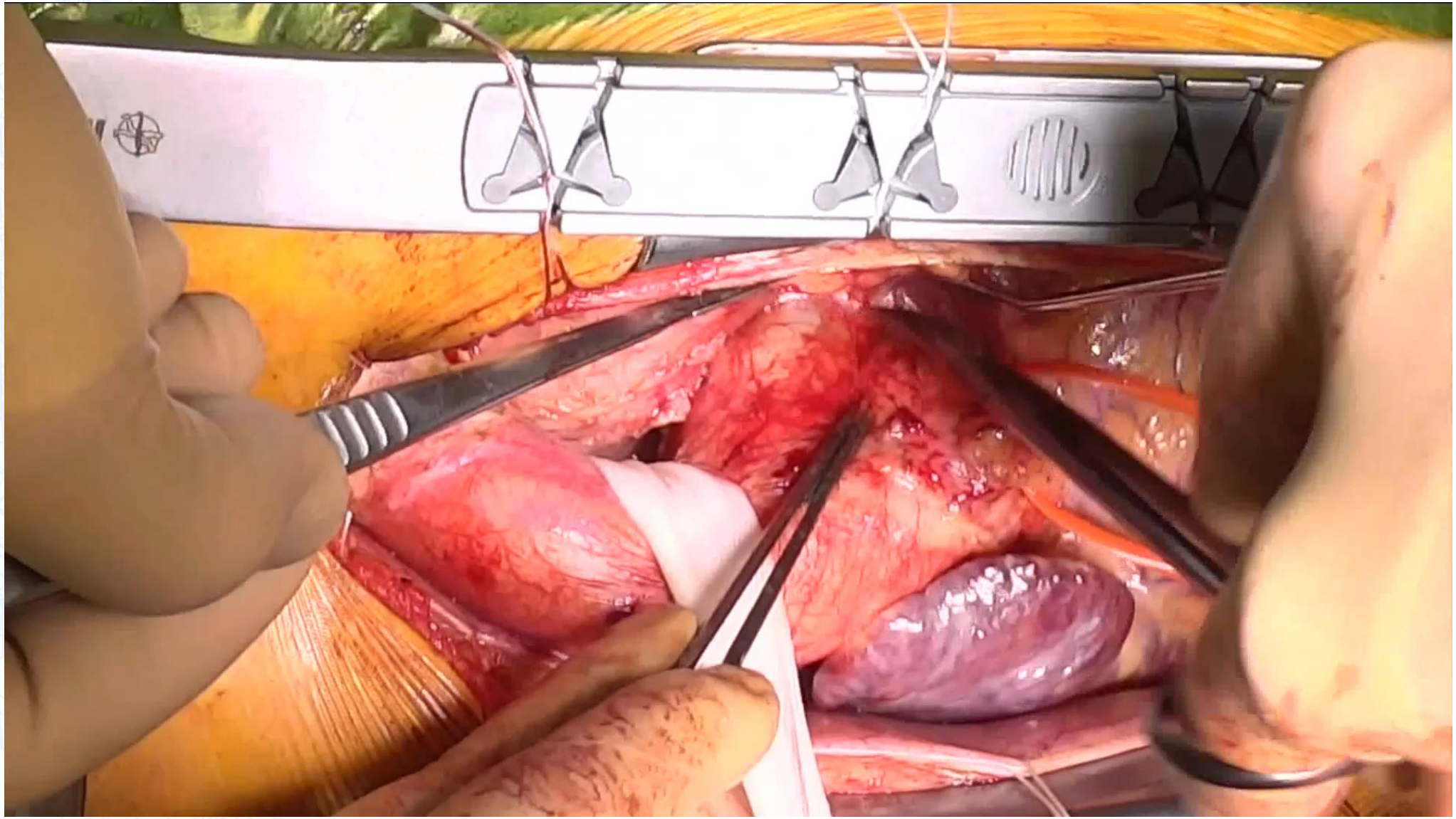
Implantace PEARS



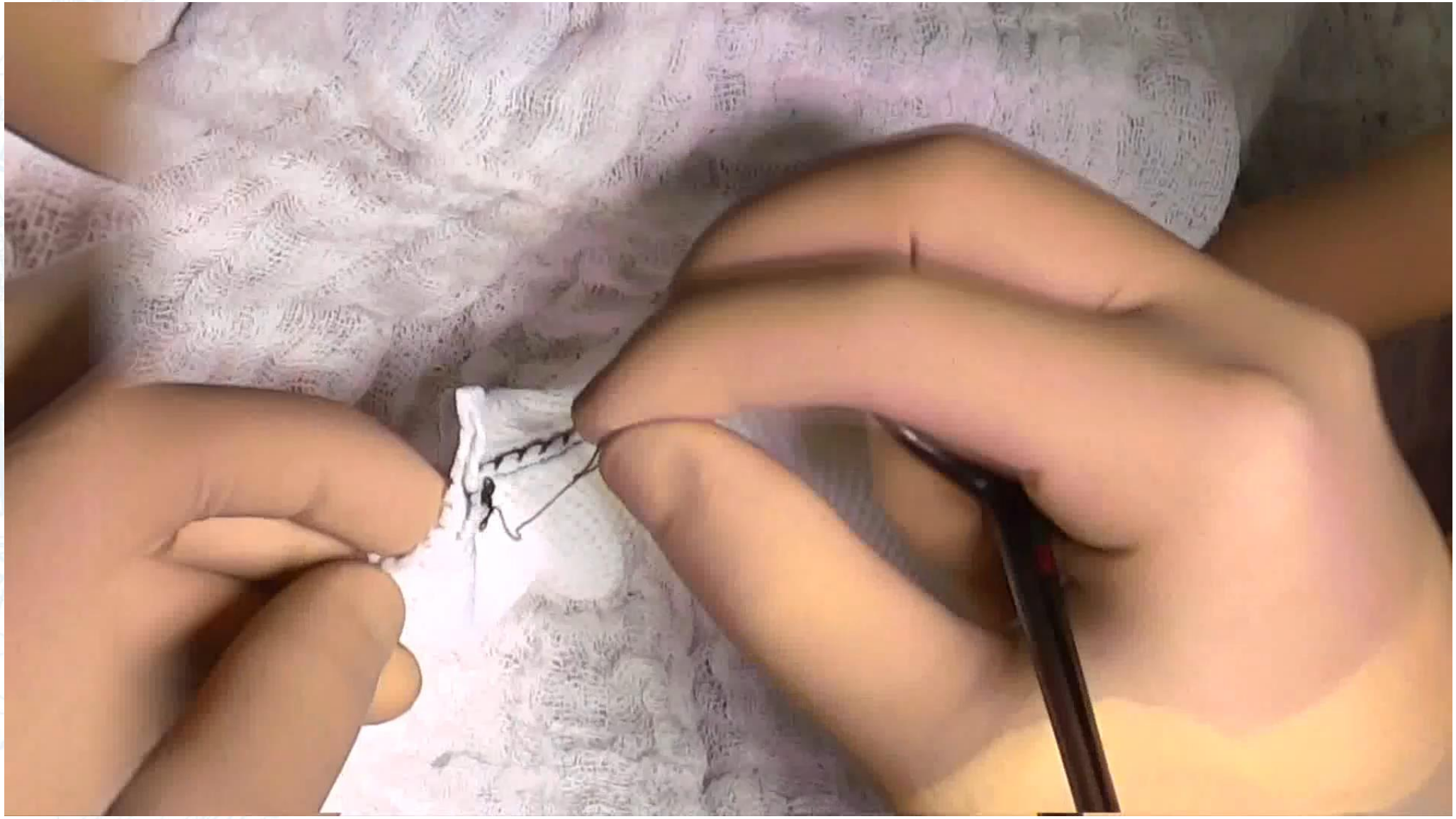
Implantace PEARS



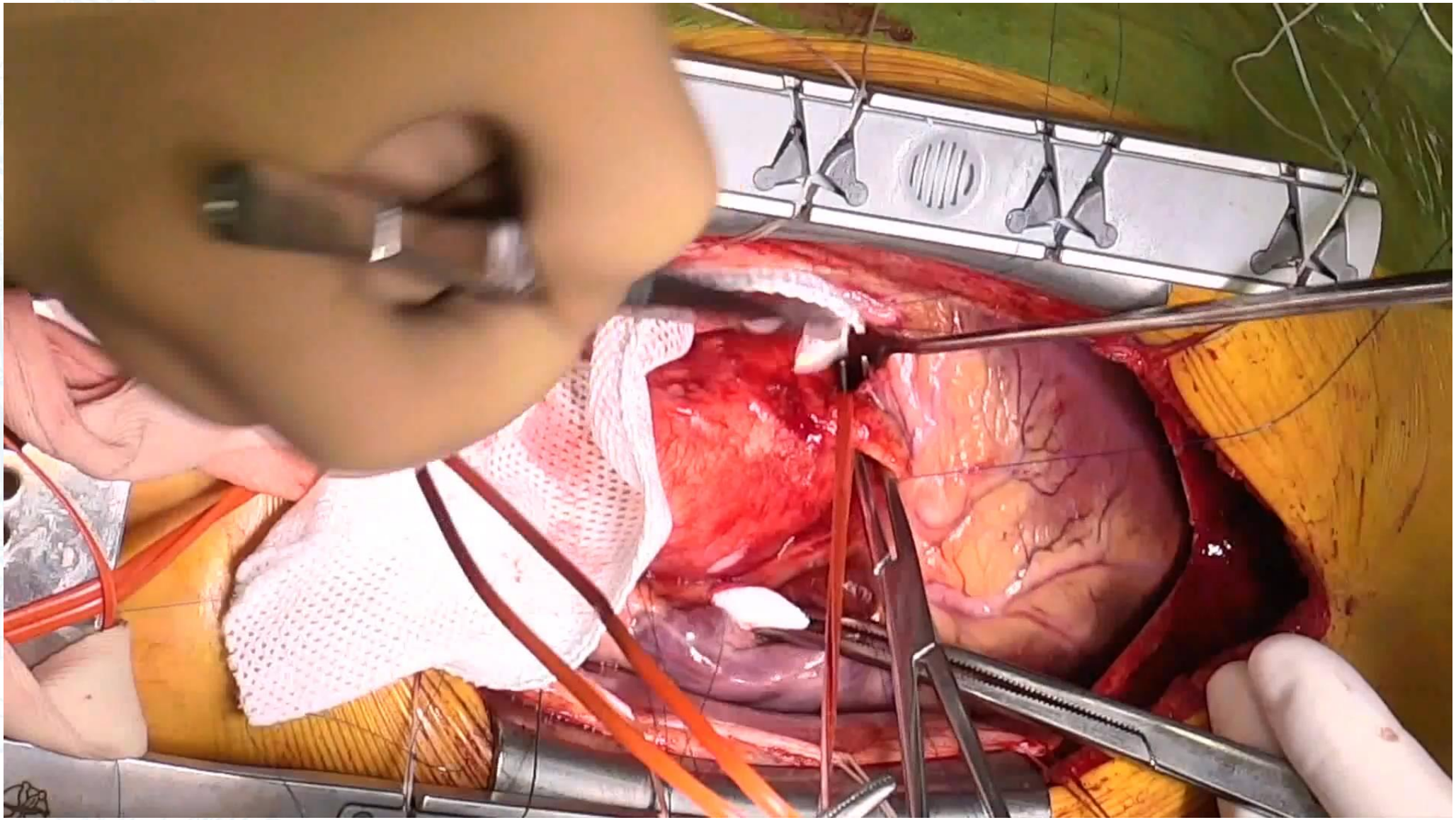
Implantace PEARS



Implantace PEARS



Implantace PEARS



PEARS – celosvětový report

01. května 2025

n = 1317 (363 žen, 956 mužů), 3-80 let

MFS, bikuspidní aortopatie, L-D sy, Turner sy, E-D sy, MYBCP3, ACTA2 mutace, TGA, Ross/PEARS

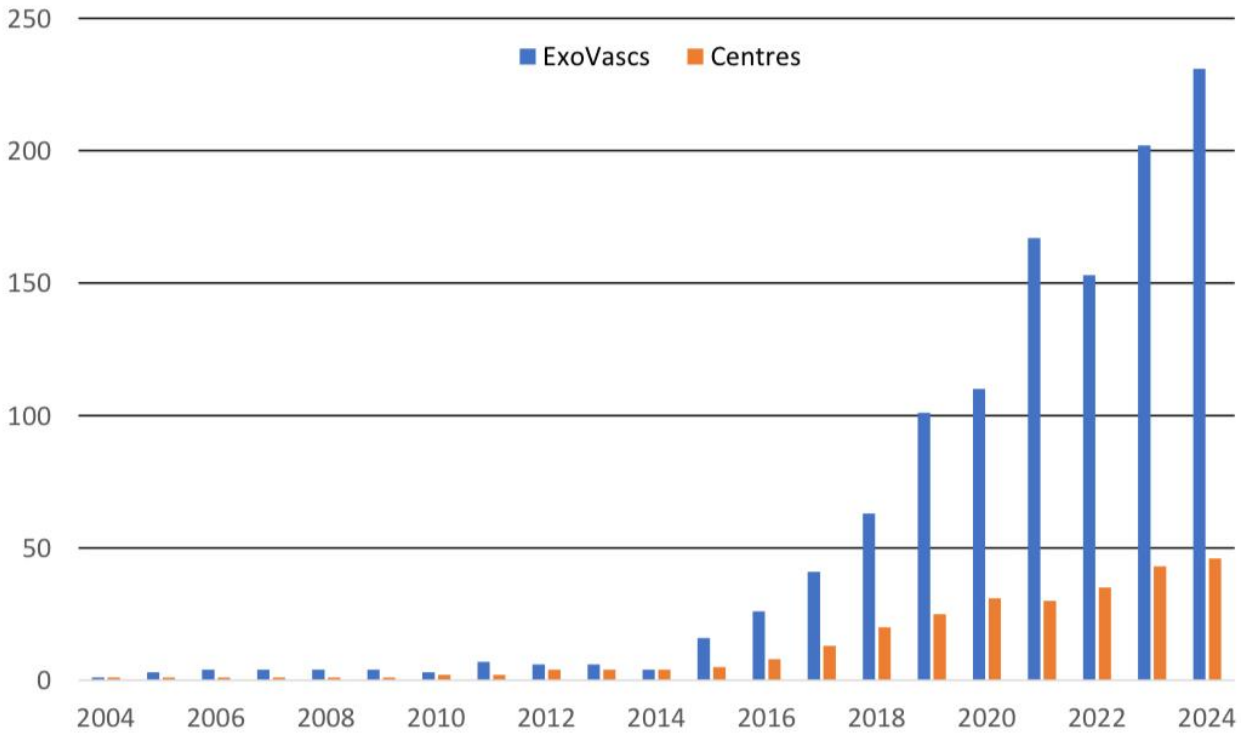
79% bez mimotělního oběhu

#1 > 20 let

50 pacientů > 10 let

319 pacientů > 5 let

14 žen - 15 narozených dětí



bez akutní disekce u implantovaných pacientů

Soubor CKTCH

03/2018-05/2025
87 implantací (9x Ross/PEARS)
19 žen; Ø věk: 48,4 (13-80let)

14x Marfanův syndrom
40x bikuspidní chlopeň
1x Turnerův syndrom
2x L-D syndrom
1x E-D syndrom
mutace MYBPC3, HYM11, …

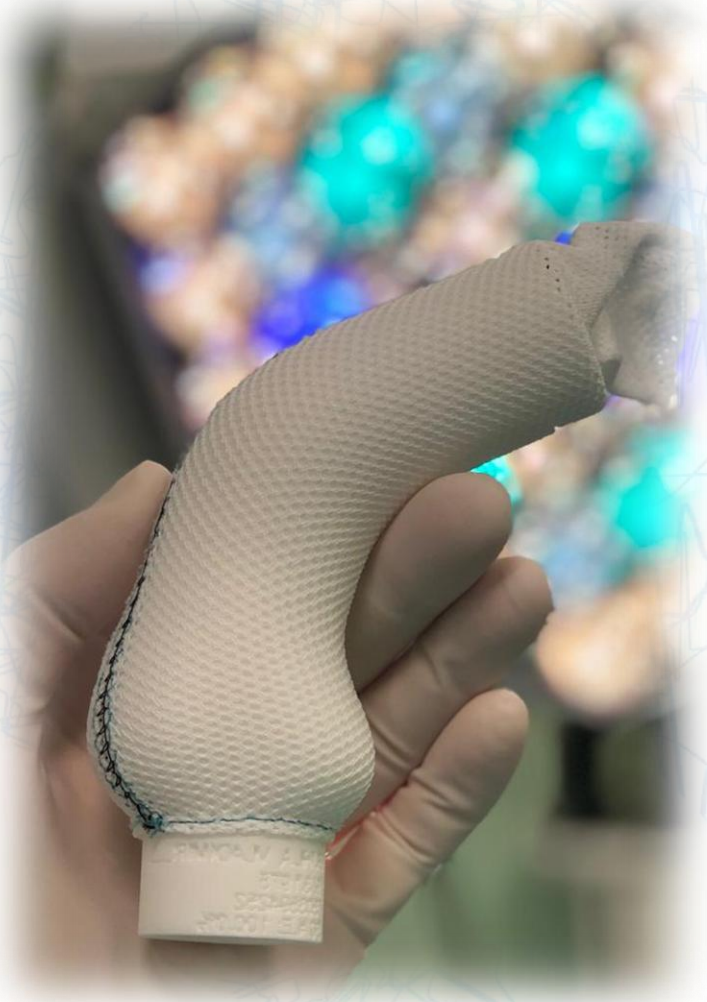
max. rozměr aorty (bulbus, AA) – Ø- 49,9mm (40-57)

78% bez ECC
8x +MVP
4x +CABG

1x reoperace – progrese AoS – SAVR bez výkonu na aortě
bez akutní disekce

nepublikovaná data

Shrnutí



chybí randomizované studie - chybí jasná opora v guidelines

- úroveň důkazů C – shoda odborníků, malé soubory, retrospektivní, registry
- stratifikace rizika

PEARS

- prevence fatální komplikace
- včasné rešení
- bez nutnosti ECC/bez zákroku na nativní chlopní
- bez antikoagulace
- zachovává endotel nativní aorty