Fallotova tetralogie
Opravena ano, vyléčena ne

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EDITORIAL

Adults with Tetralogy of Fallot -- Repaired, Yes; Cured, No

Amnon Rosenthal, M.D.

August 26, 1993
N Engl J Med 1993; 329:655-656
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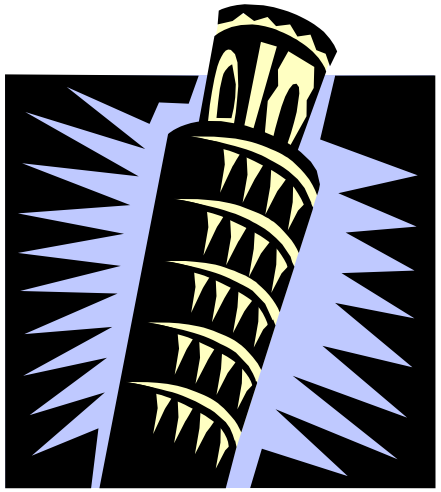
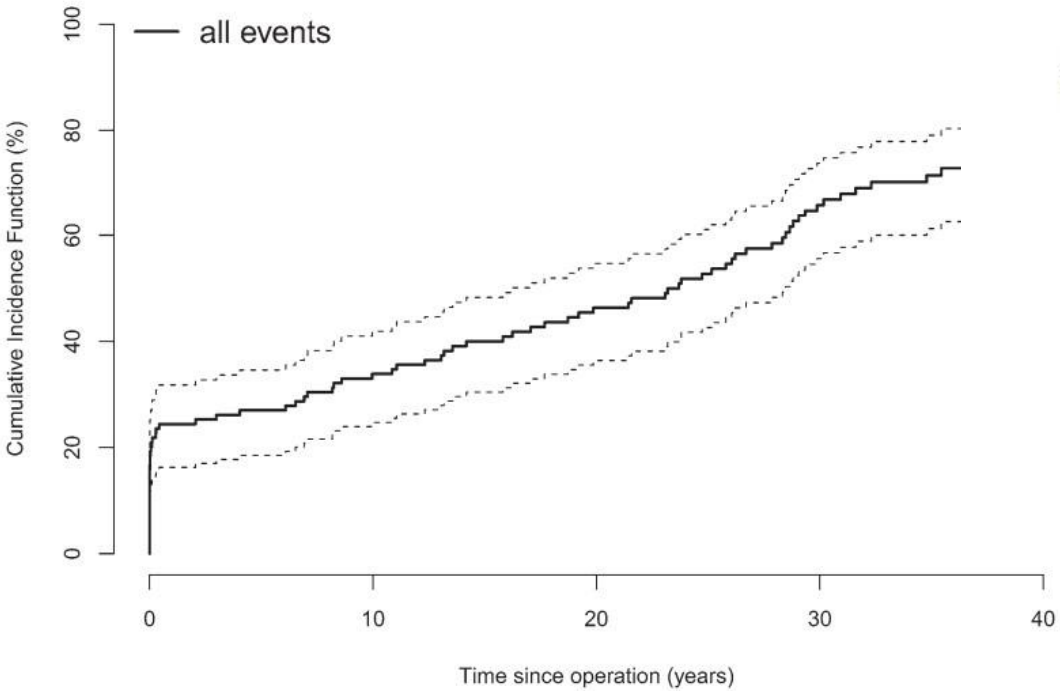
There are approximately 500,000 adults with congenital heart disease in the United States, and each year another 10,000 children who have undergone surgical repair reach adulthood[1]. The largest diagnostic category among patients undergoing repair is isolated ventricular septal defect, followed by tetralogy of Fallot. In classic tetralogy of Fallot, an anterior displacement of the infundibular septum results in a large ventricular septal defect and the development of infundibular pulmonary stenosis. Right ventricular hypertrophy is caused by right ventricular hypertension associated with both the ventricular septal defect and the pulmonary stenosis. Aortic override onto the right ventricle is the fourth . . .



Unnatural History of Tetralogy of Fallot

Prospective Follow-Up of 40 Years After Surgical Correction

Judith A.A.E. Cuypers, MD; Myrthe E. Menting, MD; Elisabeth E.M. Konings, MSc; Petra Opić, MSc; Elisabeth M.W.J. Utens, PhD; Willem A. Helbing, MD, PhD; Maarten Witsenburg, MD, PhD; Annemien E. van den Bosch, MD, PhD; Mohamed Ouhlous, MD, PhD; Ron T. van Domburg, PhD; Dimitris Rizopoulos, PhD;



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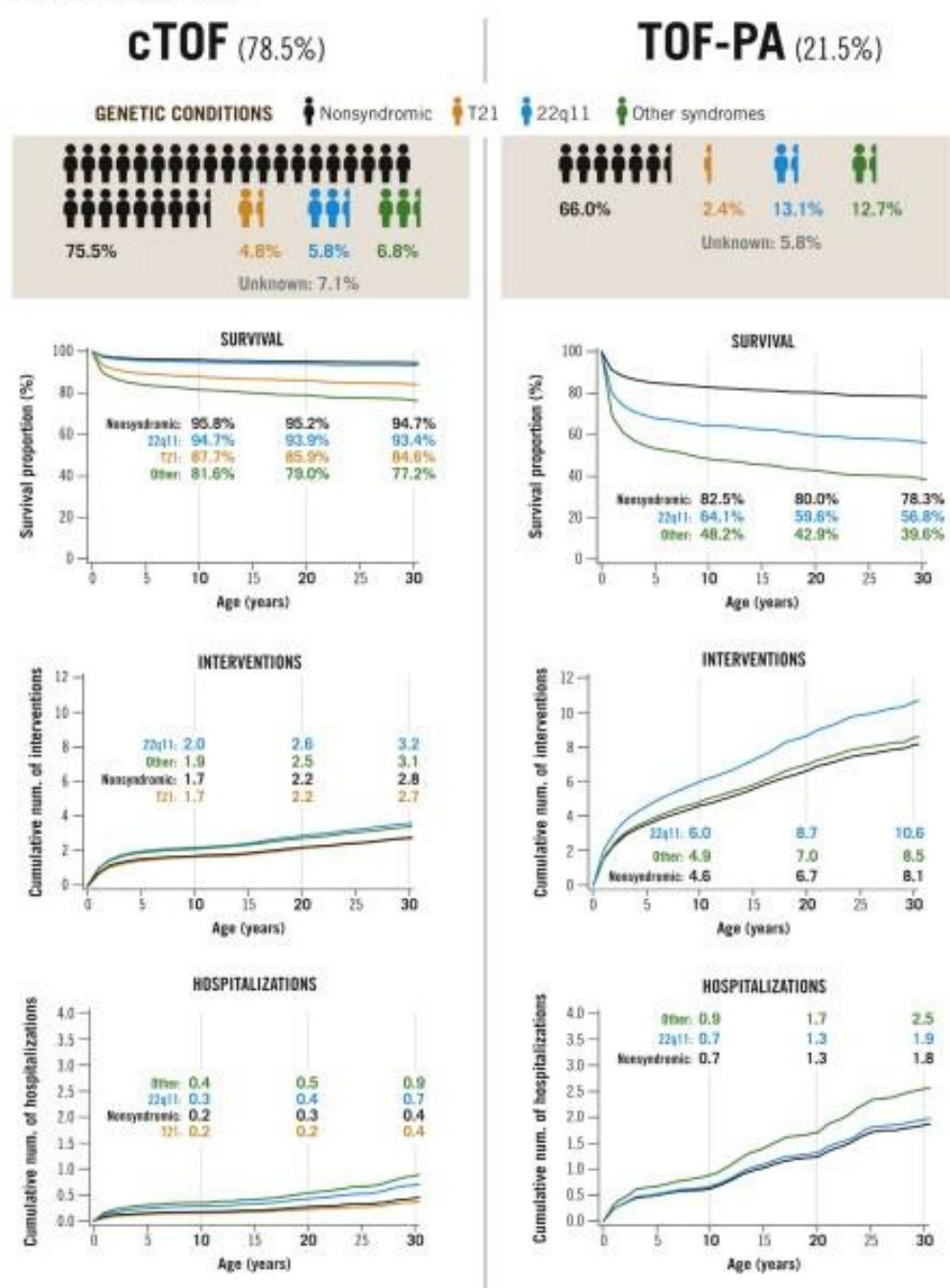
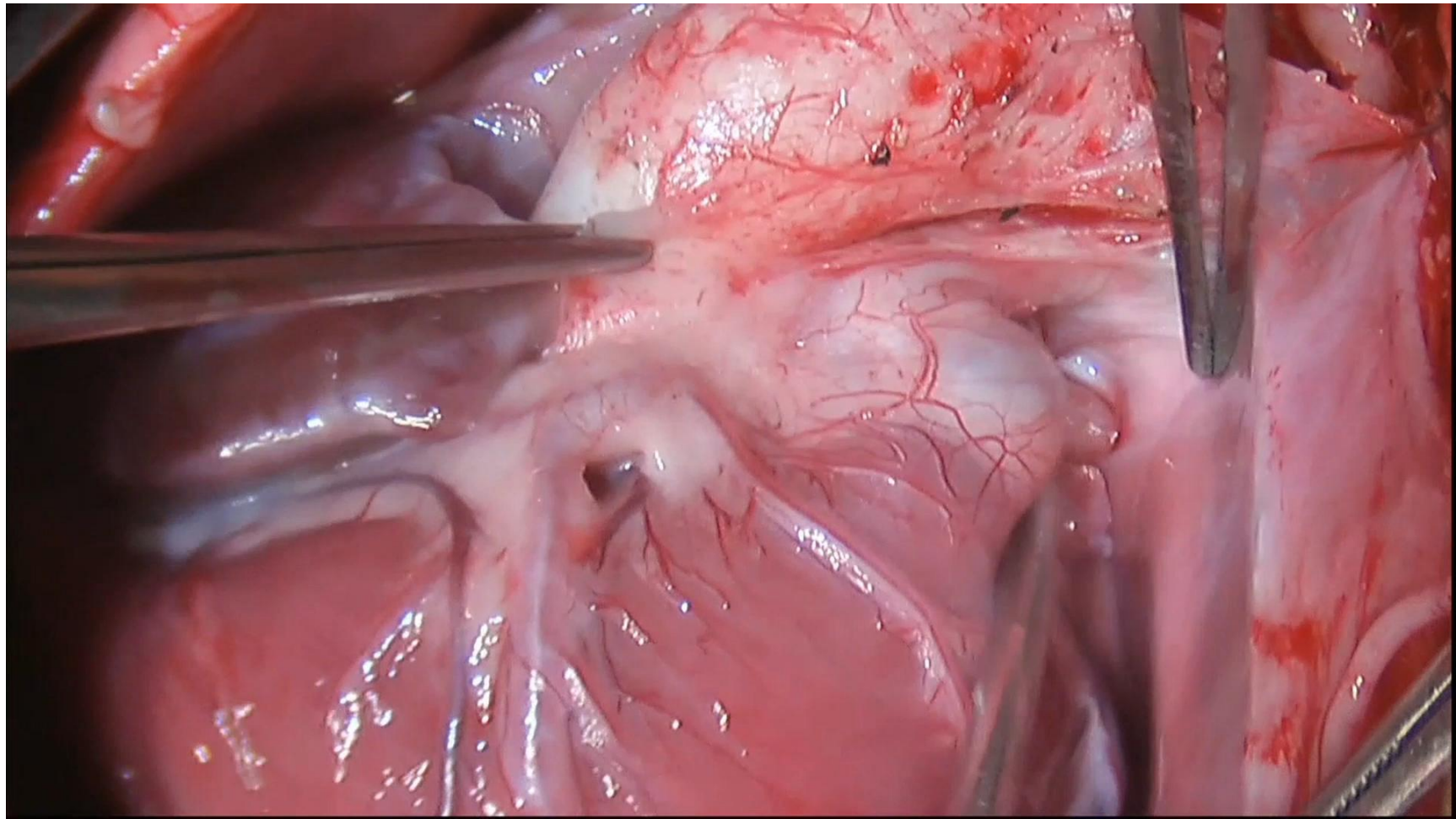


Methods

- A single nation-wide center retrospective cohort study
- 917 consecutive patients
- ToF repair: January 1979 - December 2020
  - Transatrial (TAT), transventricular (TV), transannular (TAN), transannular with monocusp (MC)
- Data from the institutional clinical database
- Cross-mapping with
  - National Death Registry
  - National Registry of Cardiovascular Interventions in Adults
- End point
  - Death (any cause)
  - Surgical or catheter reintervention
- Statistics
  - Kaplan-Meier survival probability (log-rank statistics)
  - Cox regression (mutivariable)

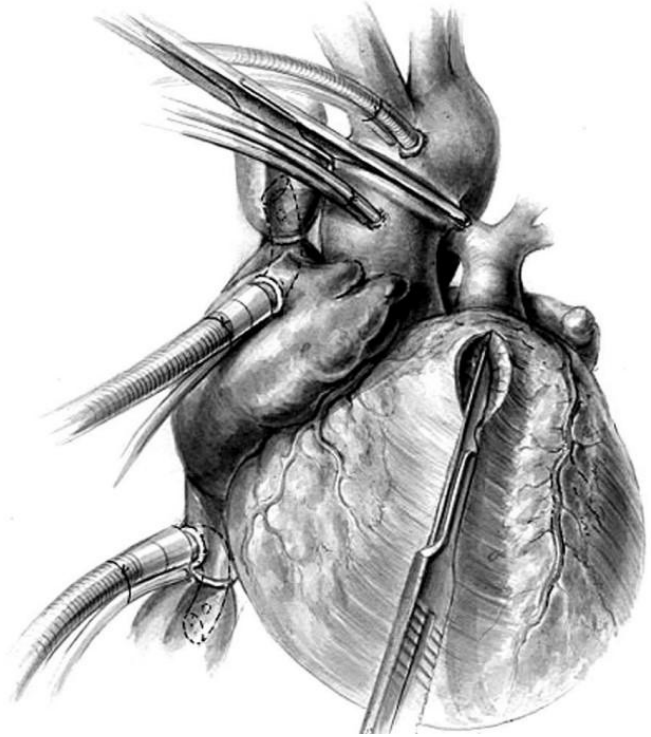
Surgical technique - video

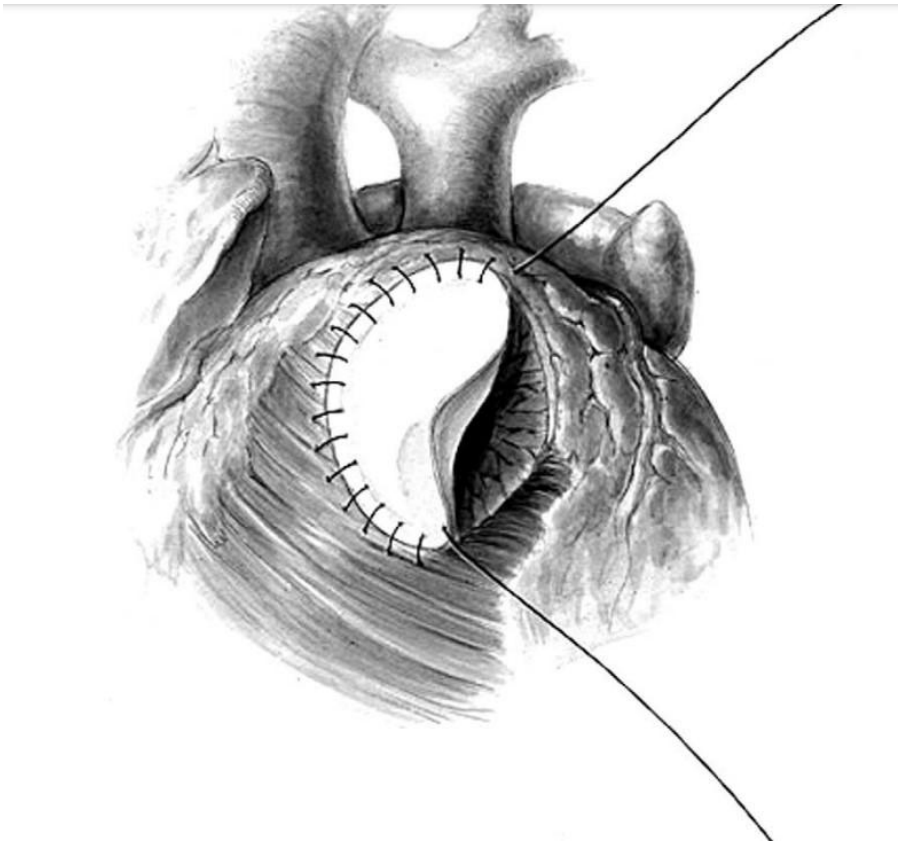
- Transatrial repair (TAT)



Surgical technique

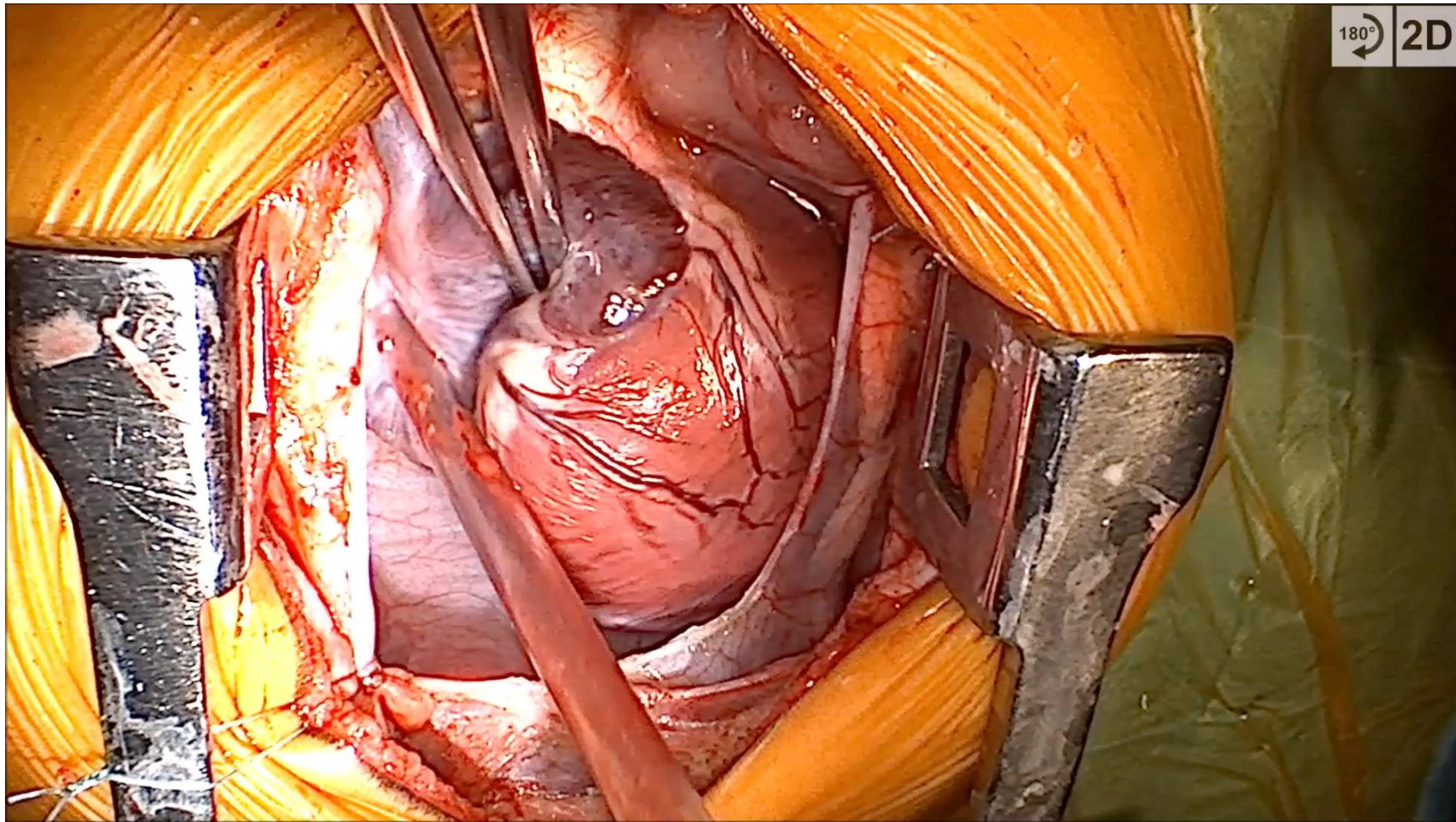
- Transventricular repair (TV)
- Schéma





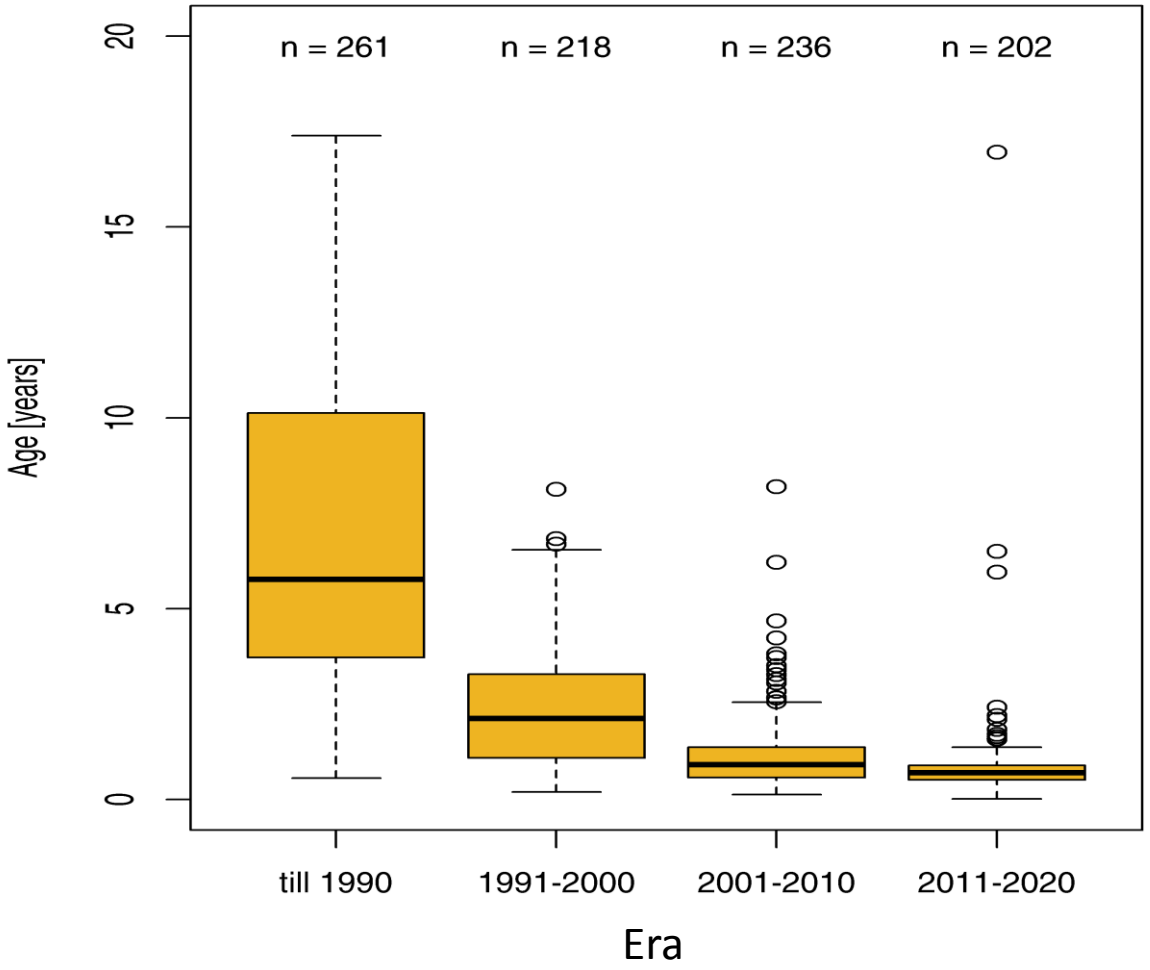
Surgical technique - video

- Transannular (TAN) +/- monocusp

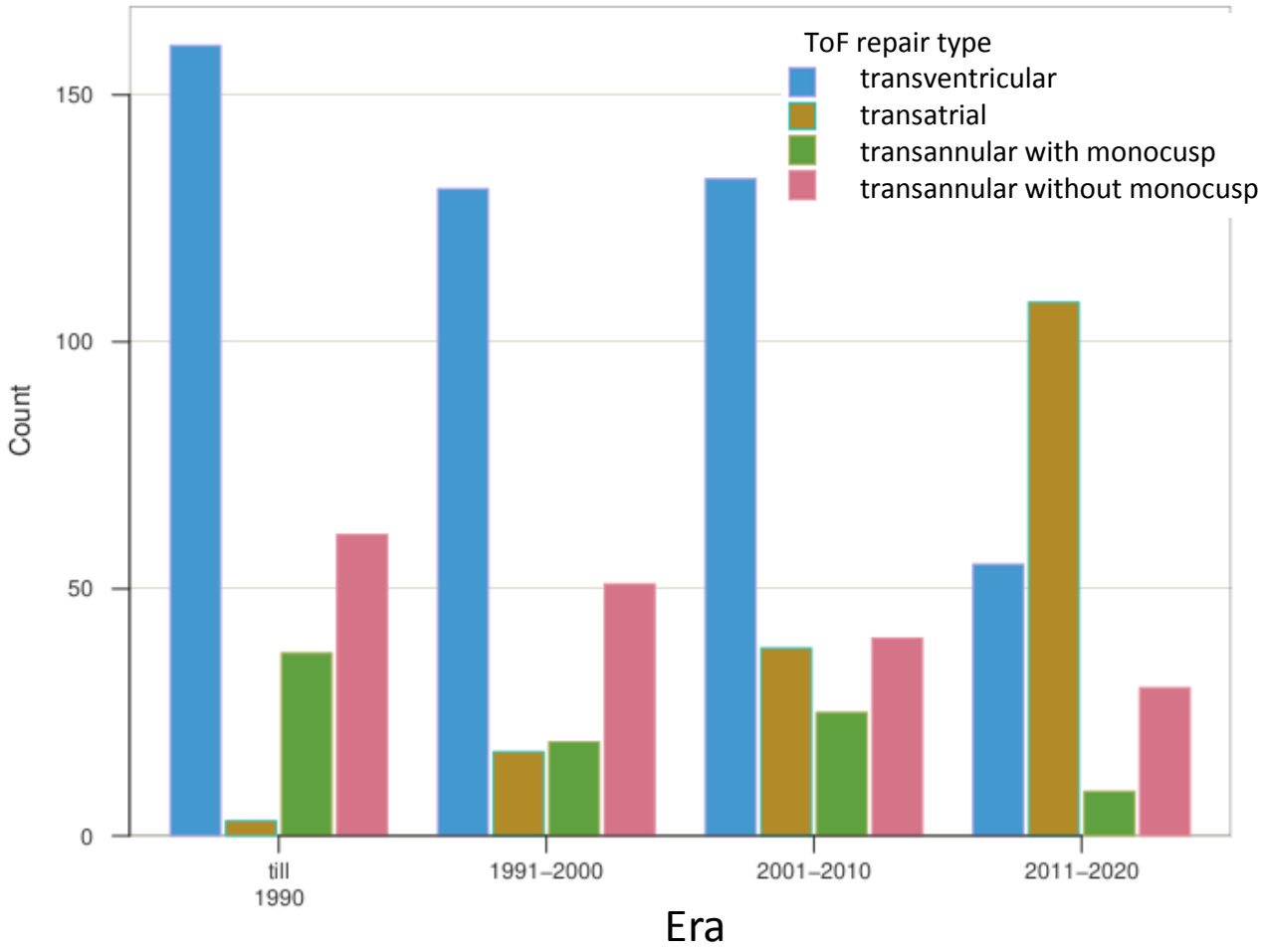


Results – age and type of initial repair

- Age at initial repair

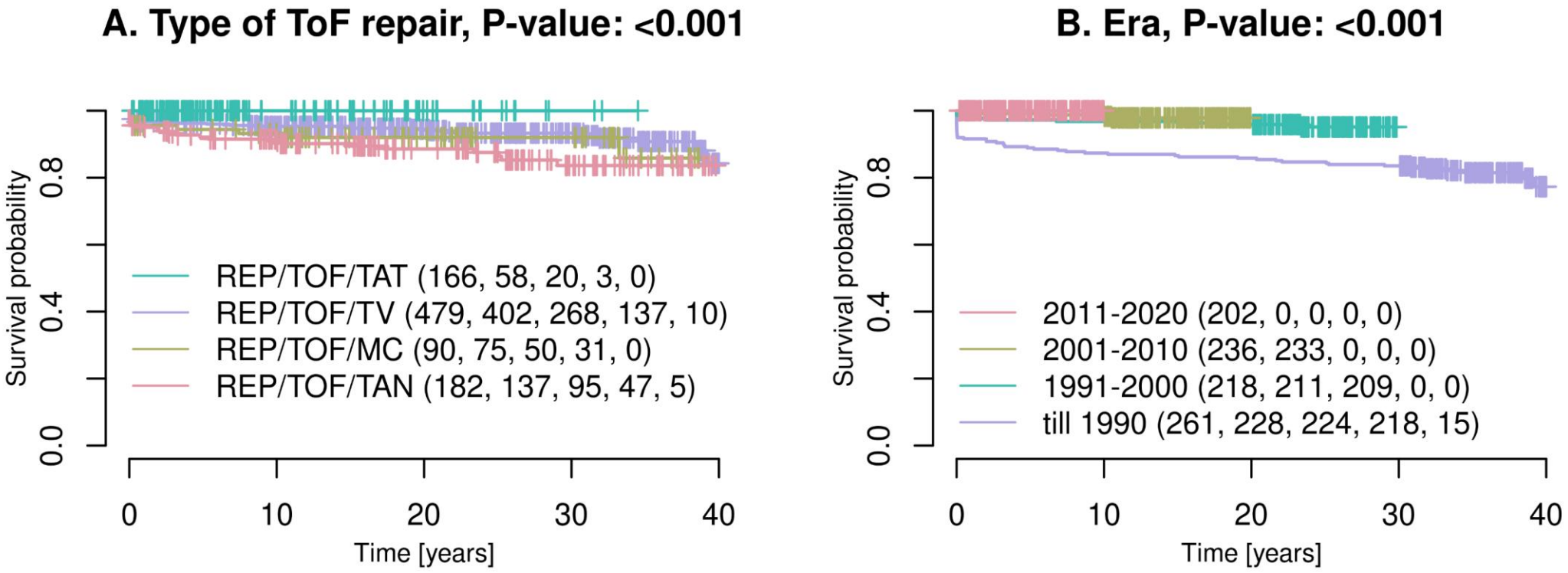


- Type of initial repair



Results: Survival

- Early mortality – 2.62 % (24/917 of patients)
  - No early death was reported since 1993
- Late mortality – 4.5 % (40/893 of patients)



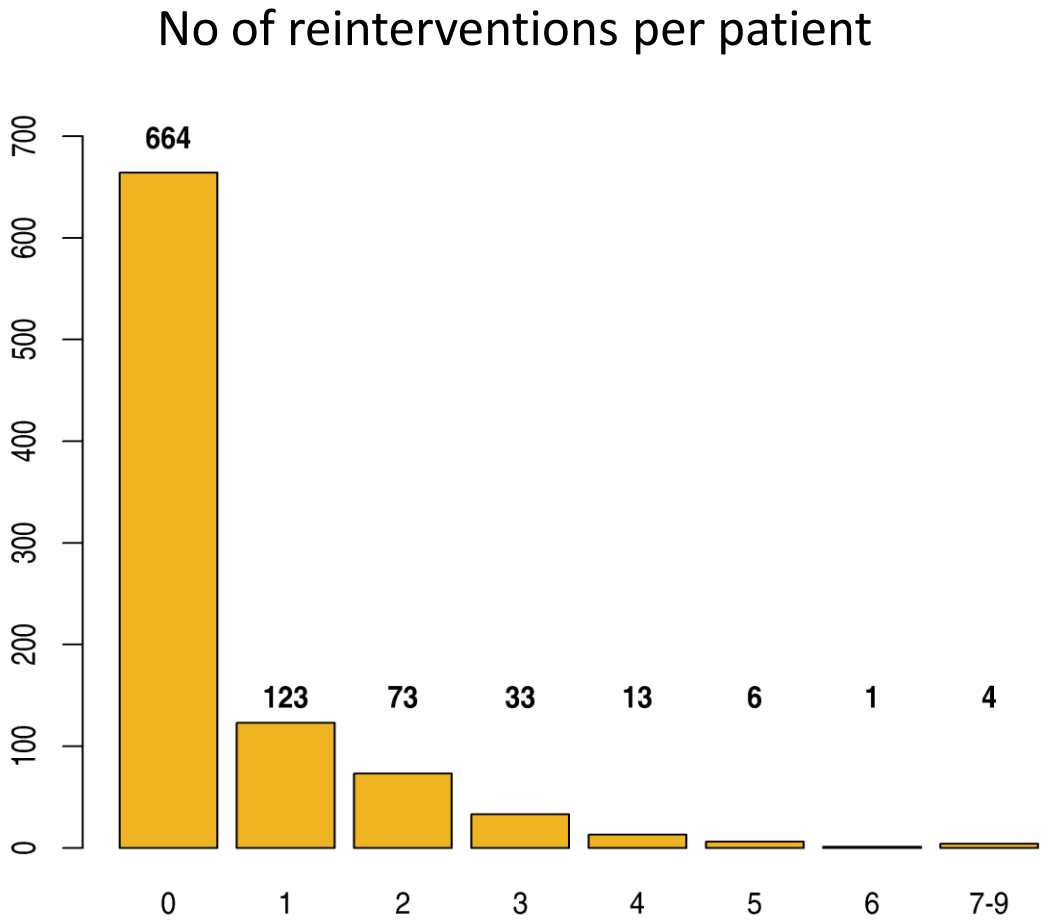
Results: Survival

Factor	HR	95%Conf.Int.	P
Surgical era			
1979 - 1990		Reference	
1991-2000	0.27	(0.12-0.59)	0.001
2001-2010	0.17	**(0.06-0.48)**	**<0.001**
2011-2020	NA		
Reinterventions			
Each additional	2,42	**(1.78-3.28)**	**<0.001**

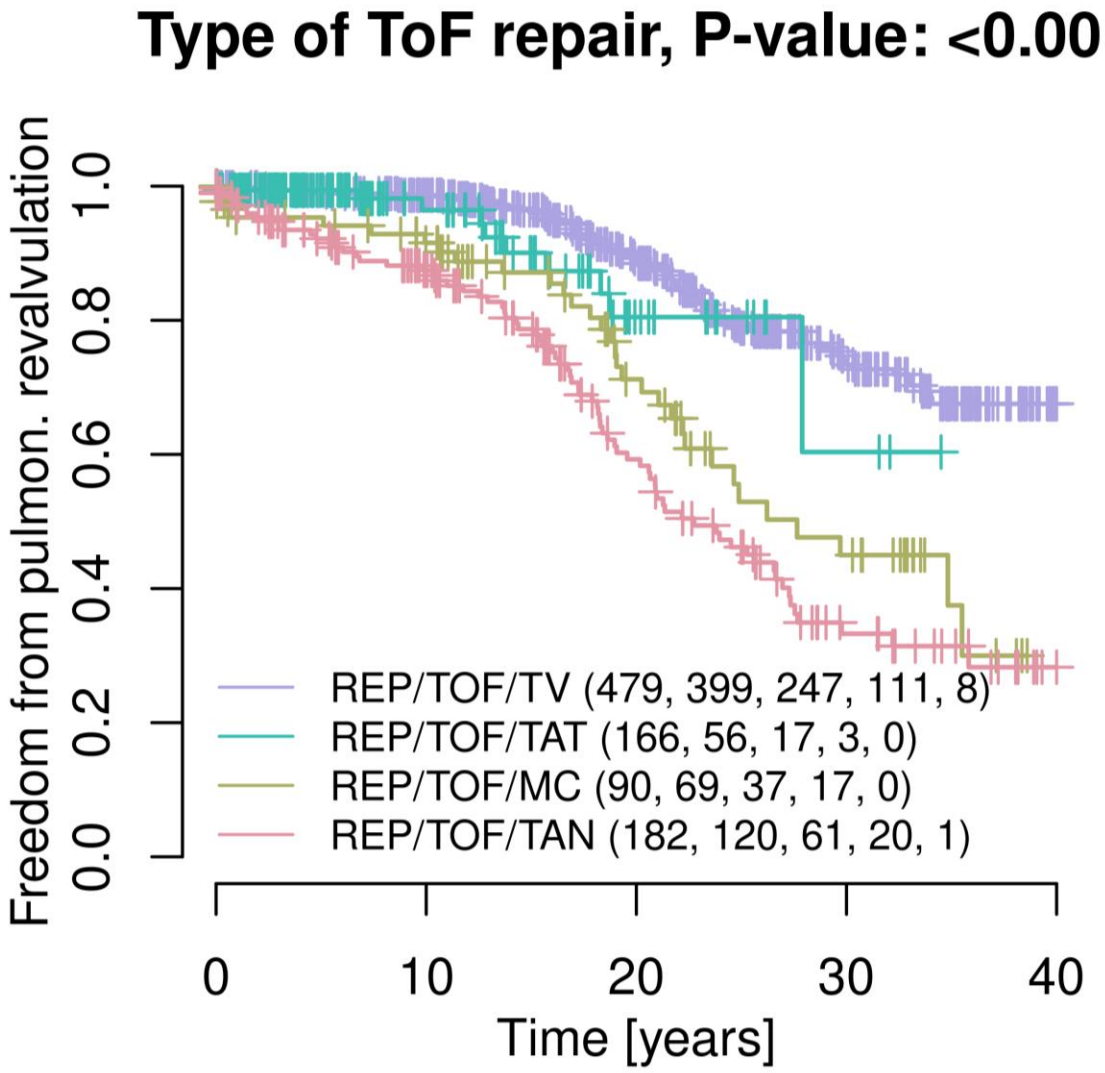
Results: Reoperations, reinterventions

- 487 surgical and transcatheter procedures in 253/917 patients (27.6%)

Type	N
Pulmonary revalvulation	196
Other pulmonary artery reinterventions	83
Right ventricular outflow tract reinterventions	35
Tricuspid valve revisions	43
Aortic valve revisions	7
Ascending aorta revisions	7
Mitral valve revisions	4
Coronary artery revisions	3
Residual VSD closure	10



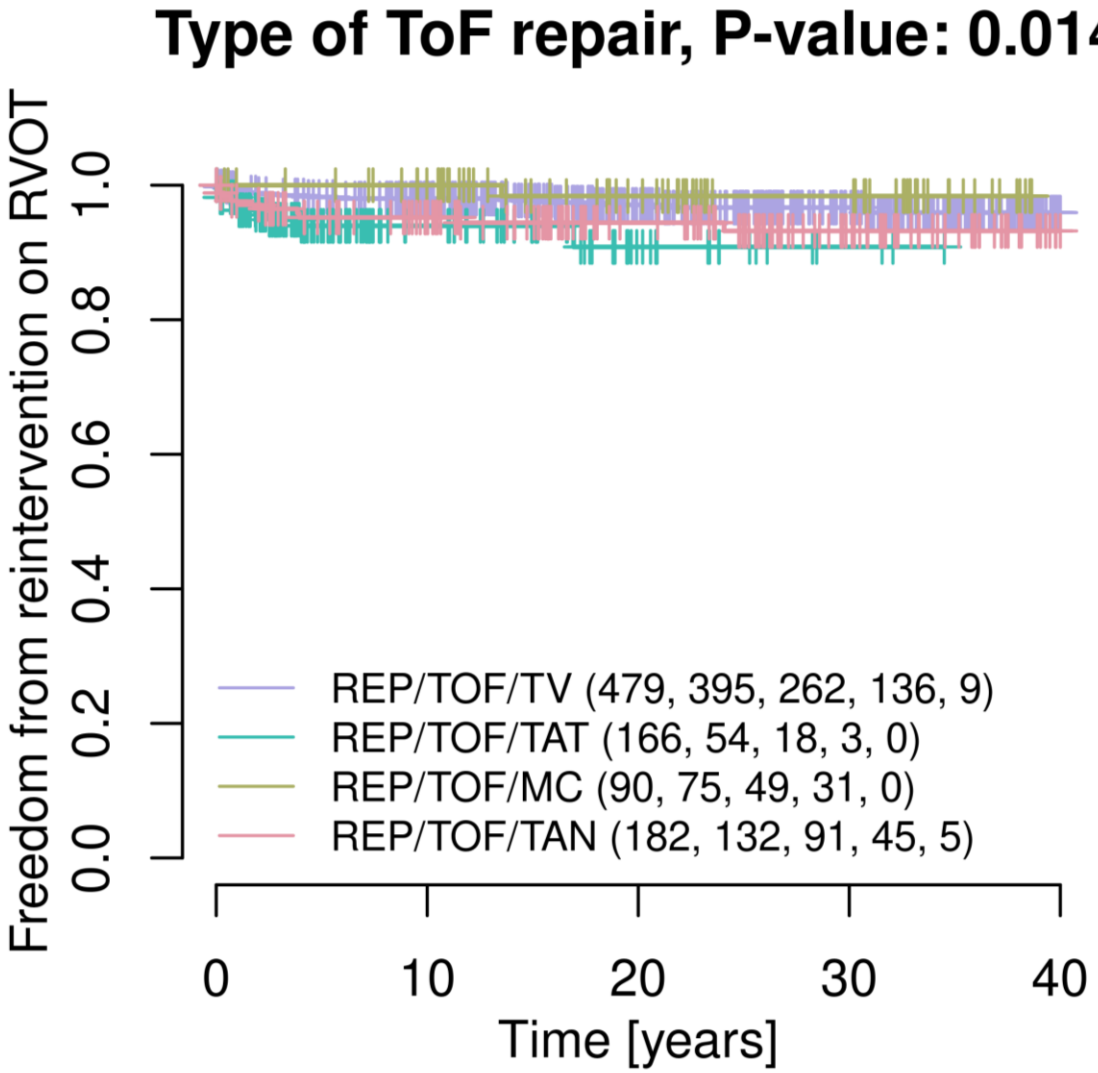
Results – Freedom from pulmonary revalvulation



Cox regression

Factor	HR	95%Conf.Int.	P
Age at initial repair			
Increase by 1 year	0.93	**(0.88-0.99)**	**0.014**
Type of initial repair			
REP/TOF/TV	1.00	Reference	
REP/TOF/TAT	1.03	(0.52-2.04)	0.923
REP/TOF/MC	2.87	**(1.88-4.39)**	**<0.001**
REP/TOF/TAN	3.78	**(2.73-5.22)**	**<0.001**

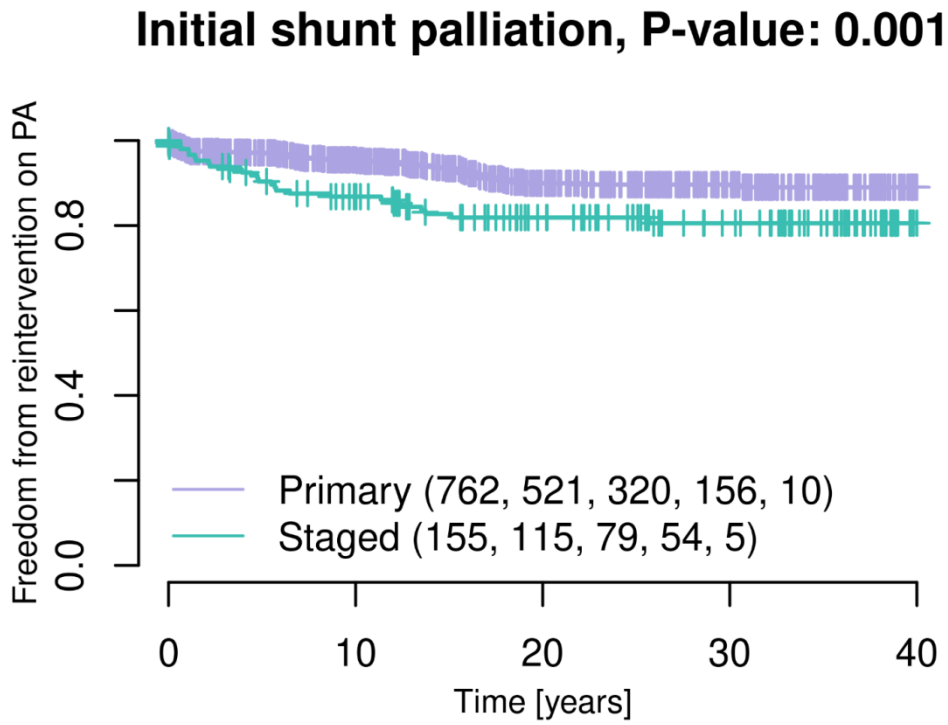
Results – Freedom RVOT reintervention



Cox regression

- None significant

Results – Freedom pulmonary artery reintervention



Cox regression

Factor	HR	95%Conf.Int.	P
Age at initial repair			
Increase by 1year	**0.61**	**(0.48-0.76)**	**<0.001**
Type of initial repair			
REP/TOF/TV	1.00	Reference	
REP/TOF/TAT	1.63	(0.77-3.49)	0.205
REP/TOF/MC	2.48	(1.28-4.79)	0.007
REP/TOF/TAN	**3.23**	**(1.91-5.44)**	**<0.001**
Initial shunt palliation			<0.001
Primary	1.00	Reference	
Staged	**4.55**	**(2.66-7.78)**	**<0.001**

Conclusions

- Most patients currently repaired in the first year of life
- Primary surgical repair is safe, approaching nowadays zero mortality
- 30-years survival >90 % in the modern era
- Burden of reinterventions, however, high
- Long-term results (specificaly pulmonary valve competence) dictated by surgical approach
- Shift to transatrial approach over time
- Specific RVOT anatomy – main driving factor for surgical decision making