Pulmonary Vein Stenosis

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DEPARTMENT OF CARDIOVASCULAR SURGERY – July 1, 2002 to June 30, 2003

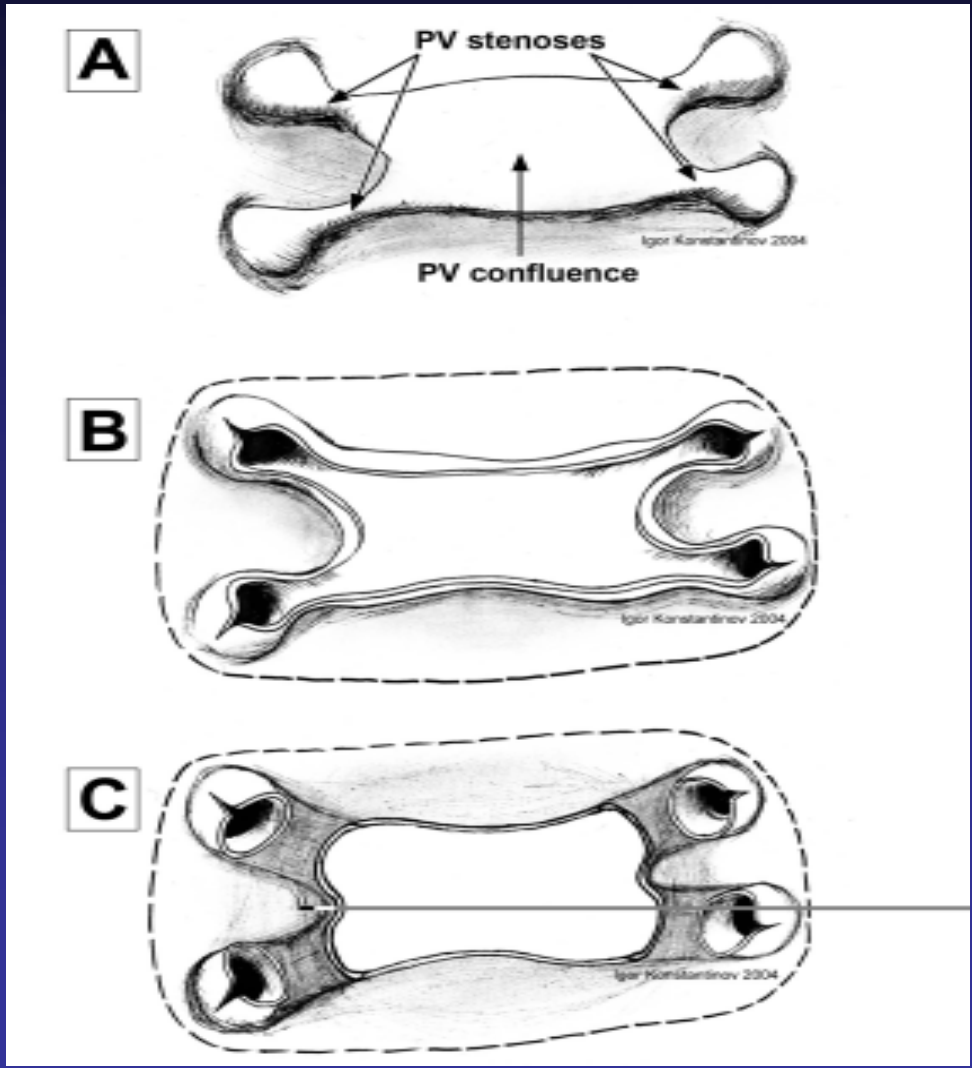
Left to Right. Dr. Tae-Jin Yun, Dr. Randall Fortuna, Dr. Julia Ritter, Dr. Nilto Carias De Oliveira, Dr. Glen Van Arsdell, Dr. John Coles, Dr. Harold Burkhart, Dr. William Williams, Dr. David Ashburn


HSC
HSC

Pulmonary Vein Stenosis

- Etiology: Acquired vs. Congenital
- Classification
- Indications for intervention
  - : For individual vein (diffuse disease, atresia..)
  - : For patients
- Surgical techniques
  - : Excision vs. Incision
    Sutureless Repair vs. Conventional technique

Excision vs. Incision



: PVS

: Incision of PV

: Excision of PV

Sutureless technique for repair of pulmonary vein stenosis

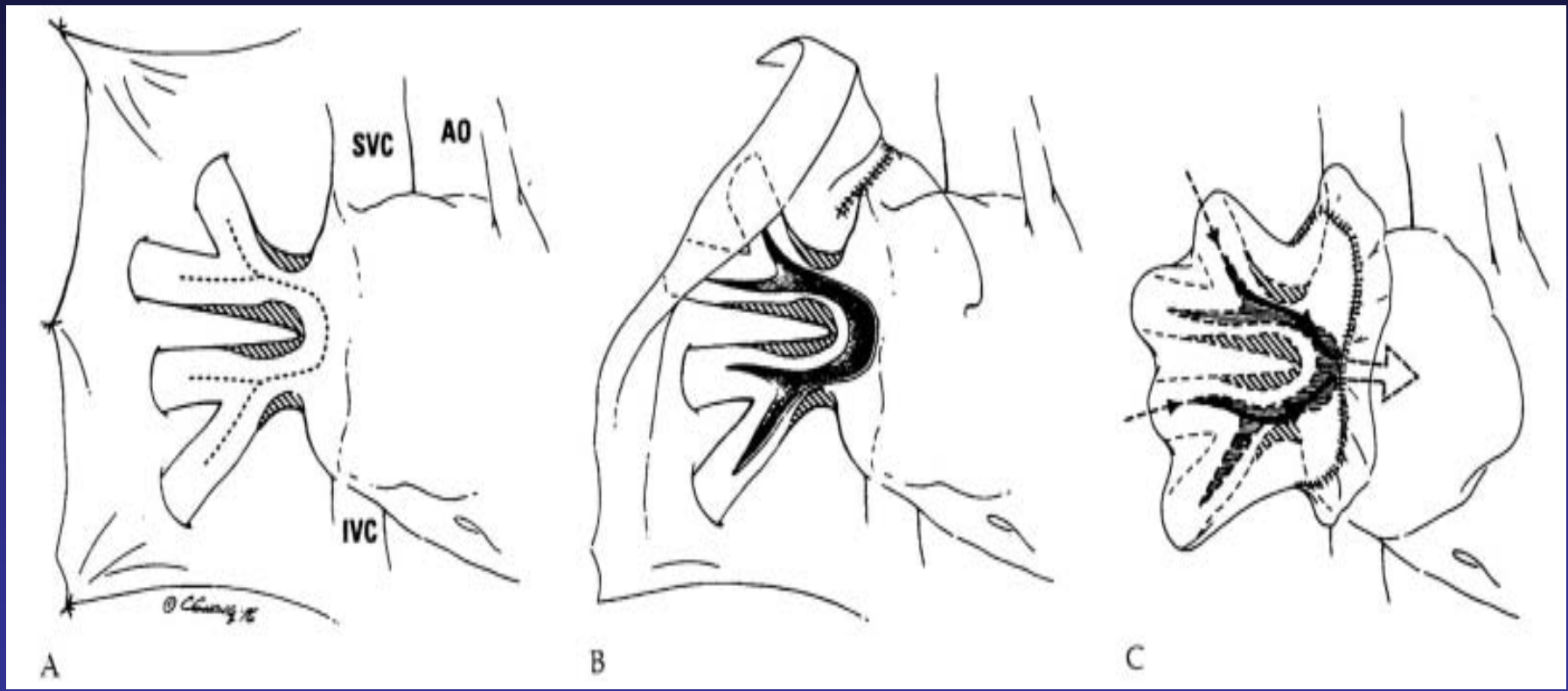
- Extension of indications -

Yun TJ, Coles JG, Konstantinov IE, Wald RM, Guerra V, Van Arsdell GS, Williams WG, Smallhorn J, Caldarone CA

The Hospital for Sick Children in Toronto


84th Annual Meeting

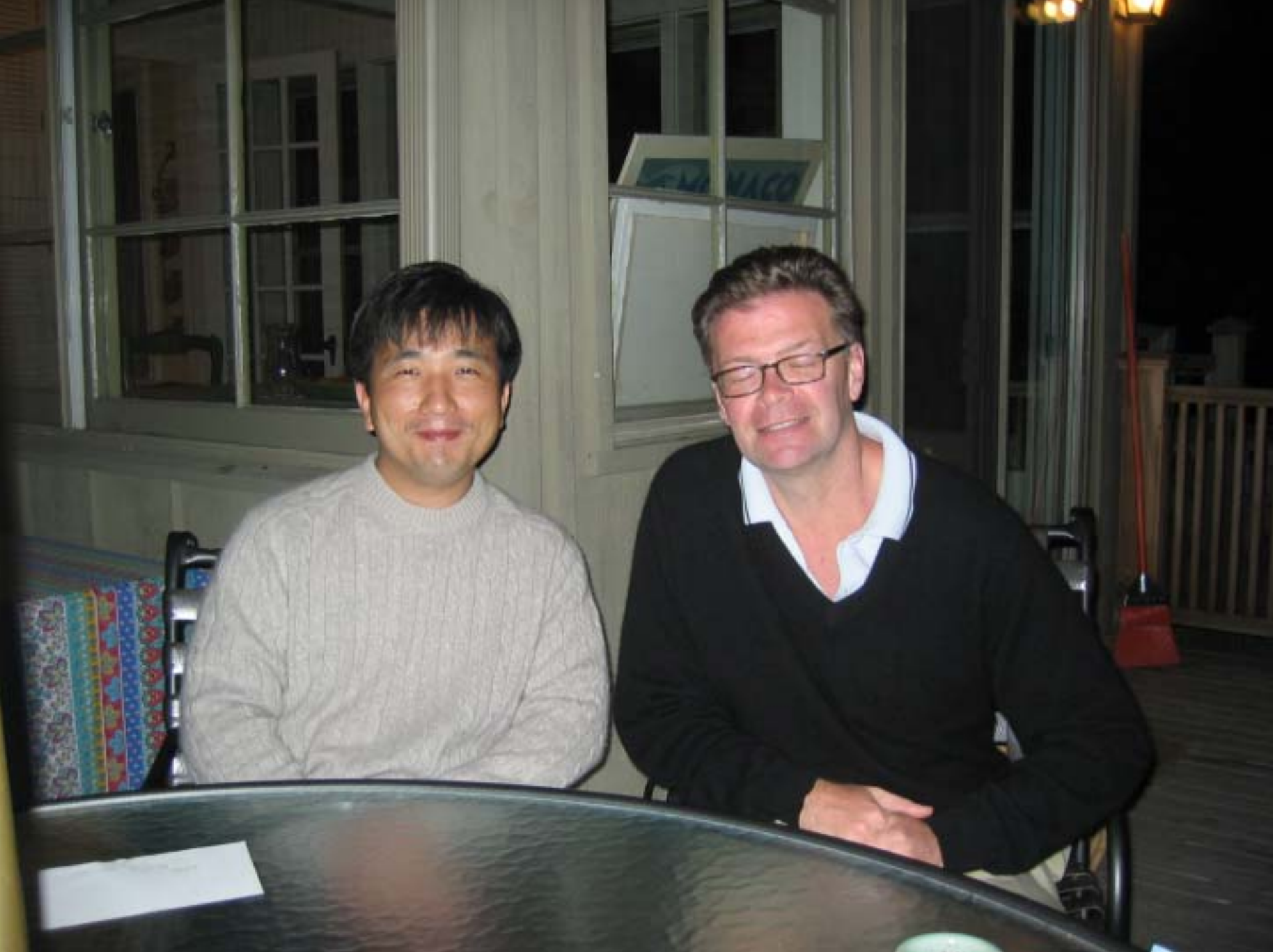
Sutureless Repair



Sutureless repair (SR)

- Theoretical advantage -

- **Avoid geometric distortion**
- **No suture material on PV**
- **Avoid restriction of ostial growth**



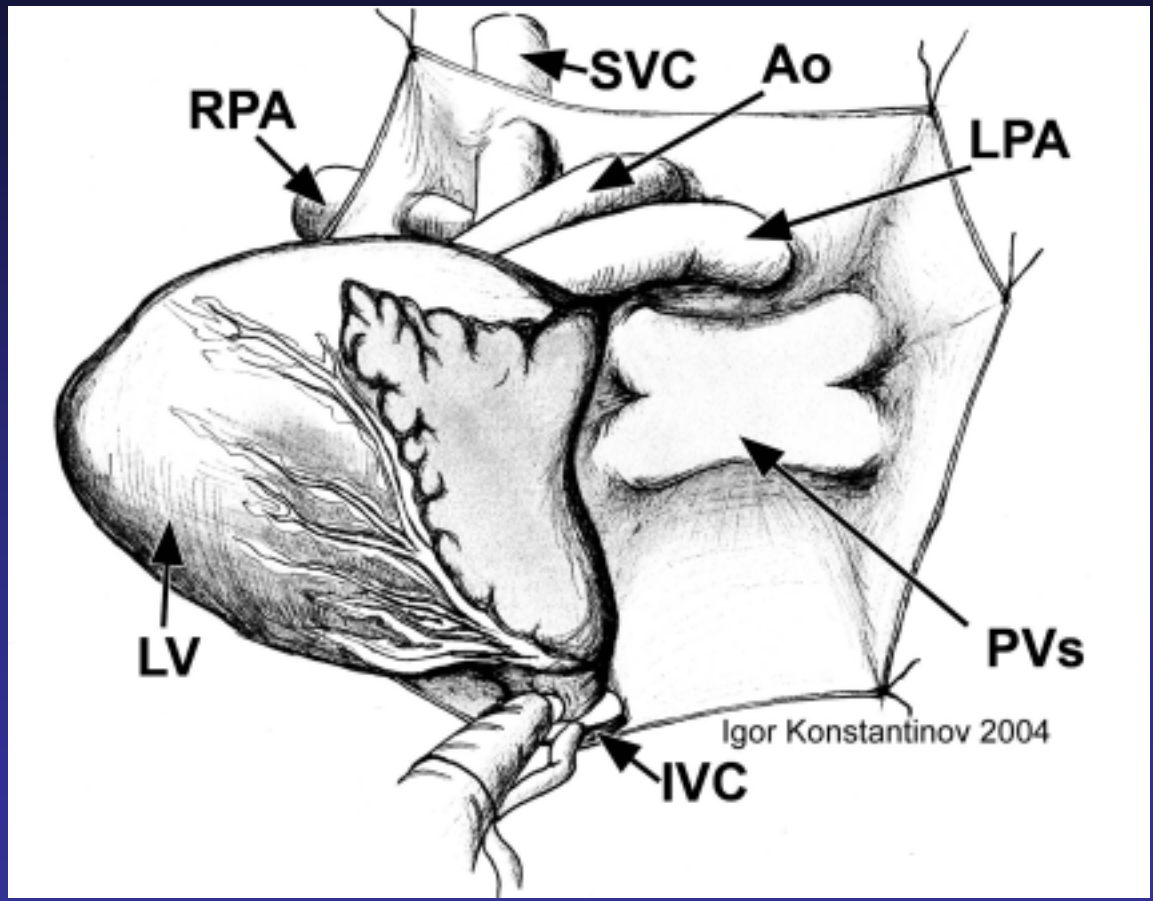

APOPTOSIS

Objectives

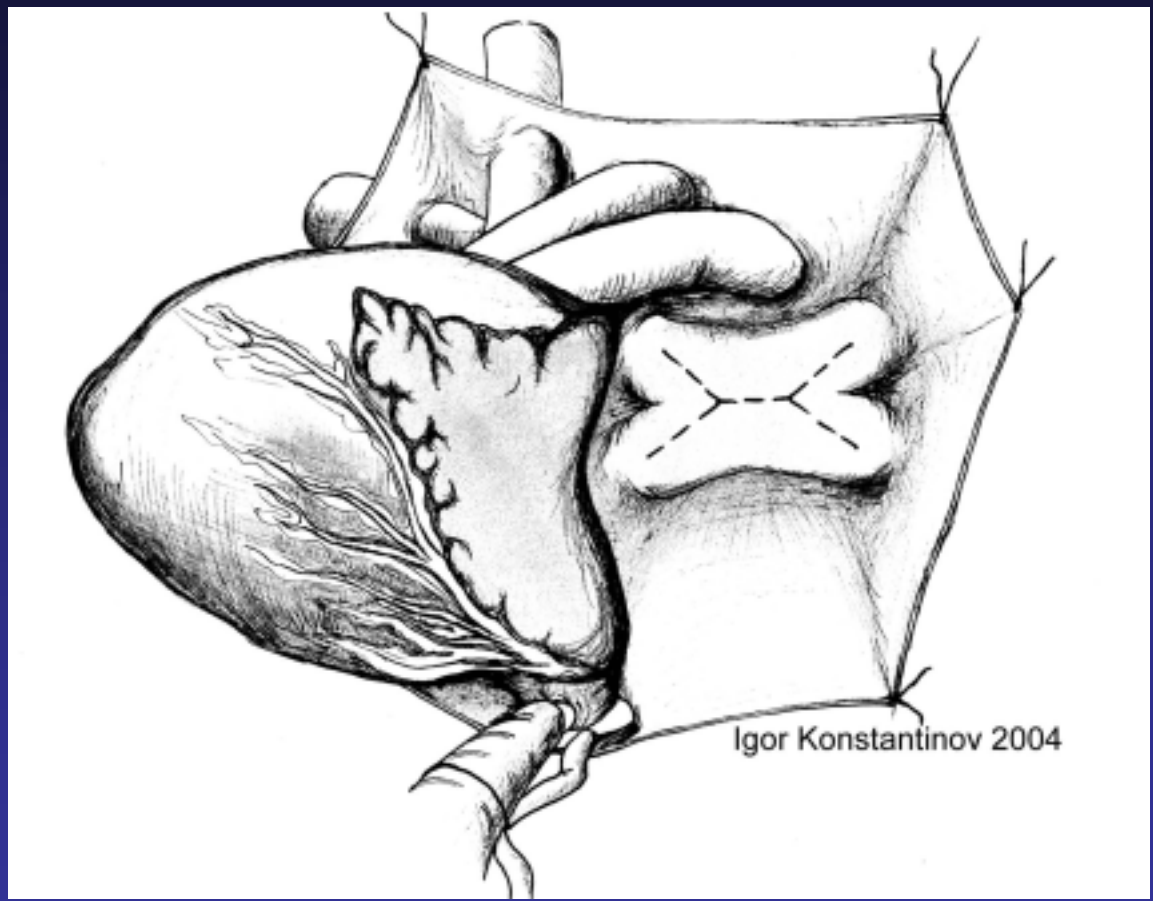
Evaluate Sutureless Repair (SR):

- **Post-repair PVS (After repair of TAPVD)**
- **Efficacy of SR for other indications**
- **Safety of SR without retrocardiac adhesion**

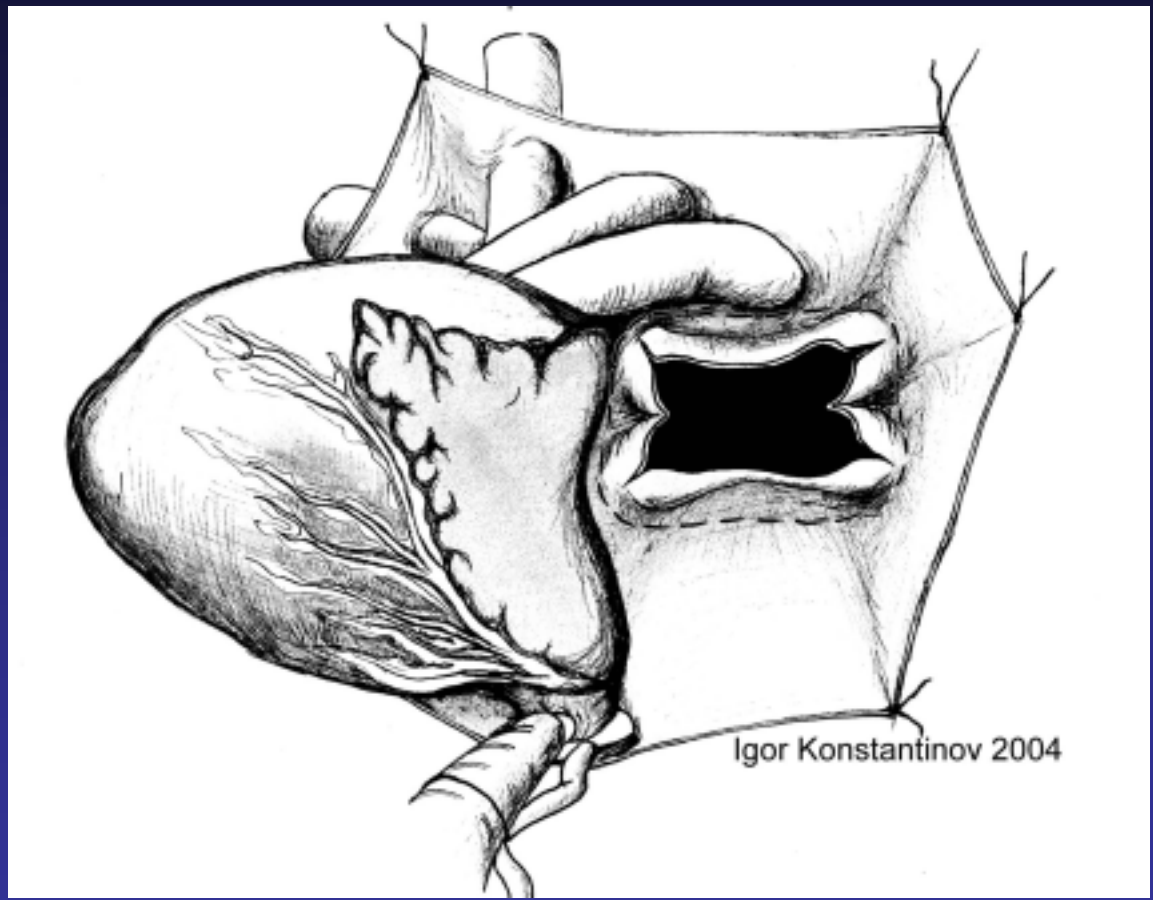
SR without Retro-cardiac adhesion



SR without Retro-cardiac adhesion



SR without Retro-cardiac adhesion



Methods

60 PVS patients over 20 years
73 procedures (40 Sutureless Repairs)
Age: 7 d – 38 m (4.4 m)

Follow-up: 1 m – 18.6 yrs (2.9 yrs)
Complete: 88%

Statistical Analysis

Retrospective analysis

Cox Proportional Hazard Survival Model

Endpoint: Re-operation or death

Variables:

- **Age**
- **Indication category**
- **Extent of disease (PVS score)**
- **Heterotaxy syndrome**
- **Types of intervention**
- **Previous cardiac operation**

Model validation with bootstrap analysis

PVS score

Individual pulmonary vein stenosis graded

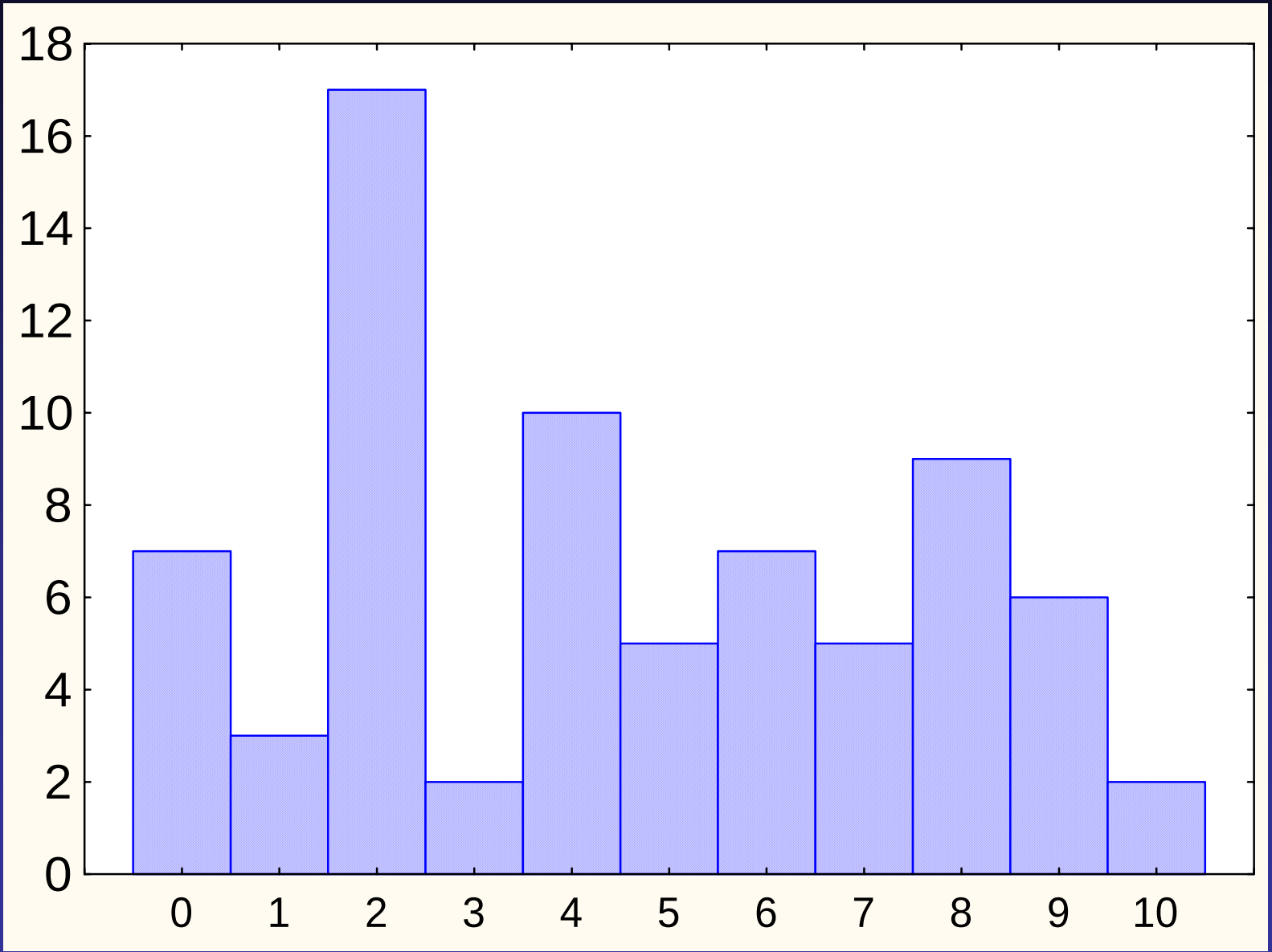
0: No PVS
1: Mild-Moderate PVS
2: Severe PVS
3: Obstruction

Summation of four PV grades

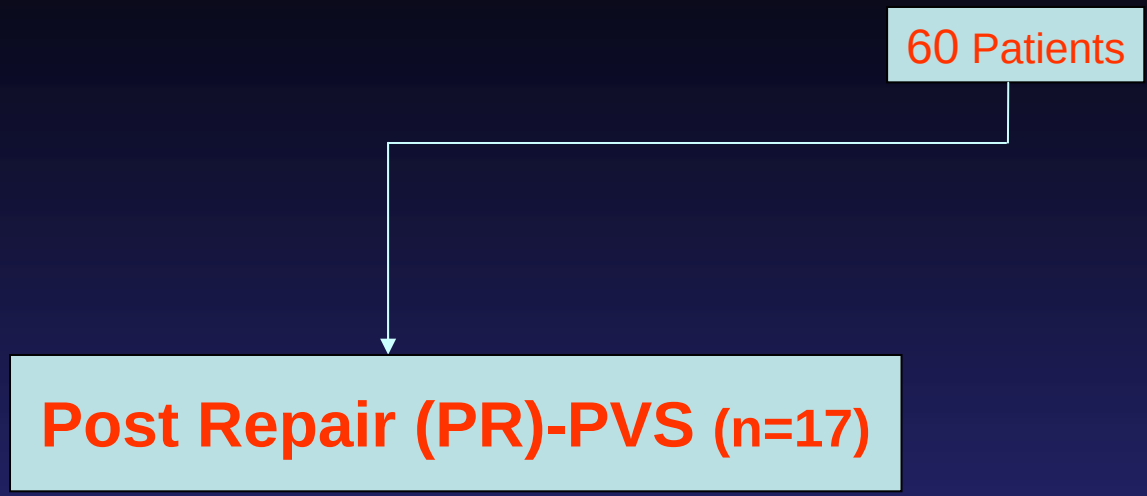
>>>> ‘PVS score’

Possible range: 0 - 12

PVS score



60 Patients

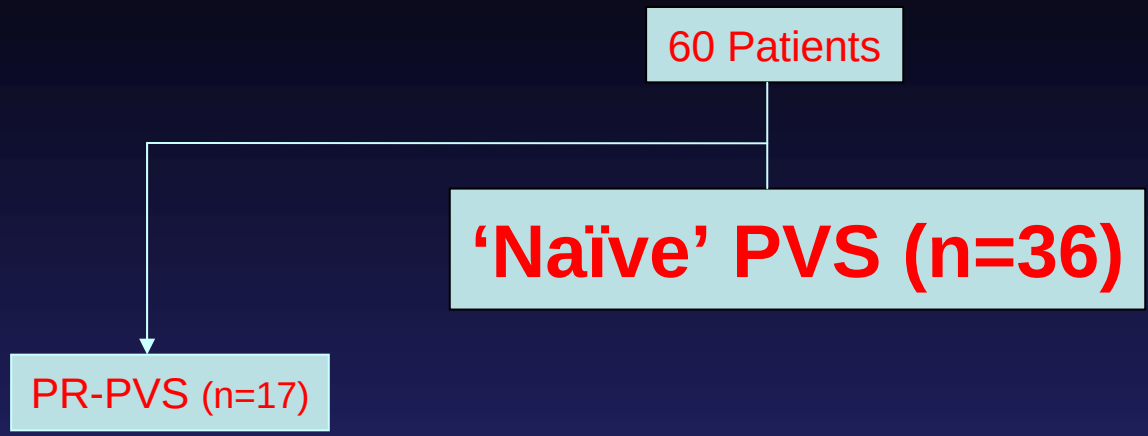

60 Patients
Post Repair (PR)-PVS (n=17)

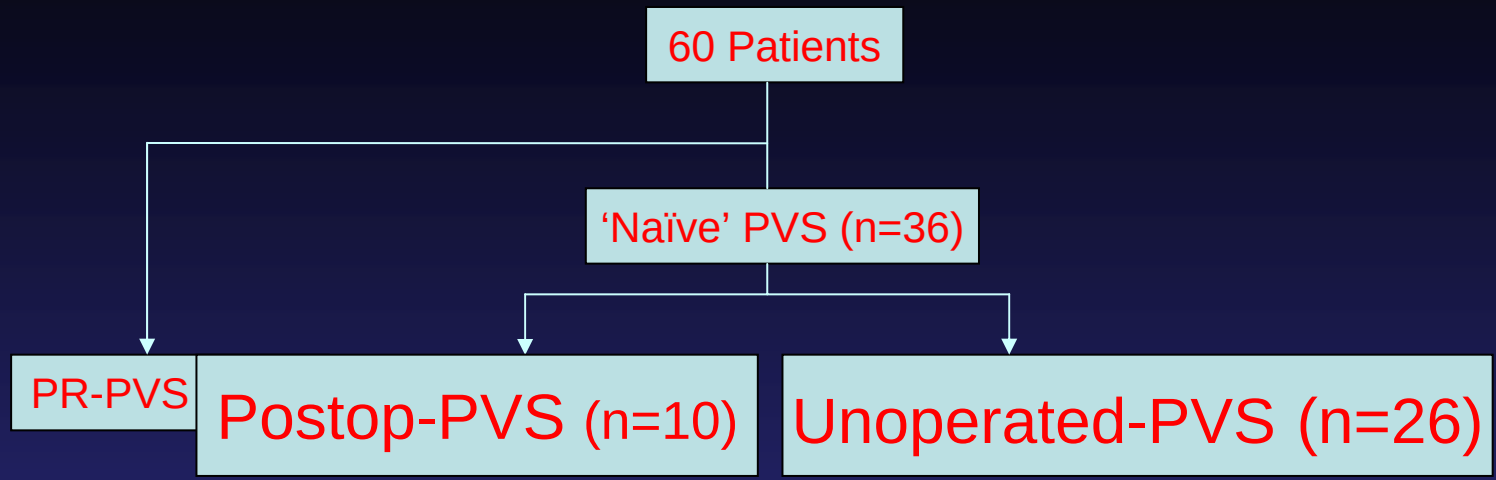
60 Patients

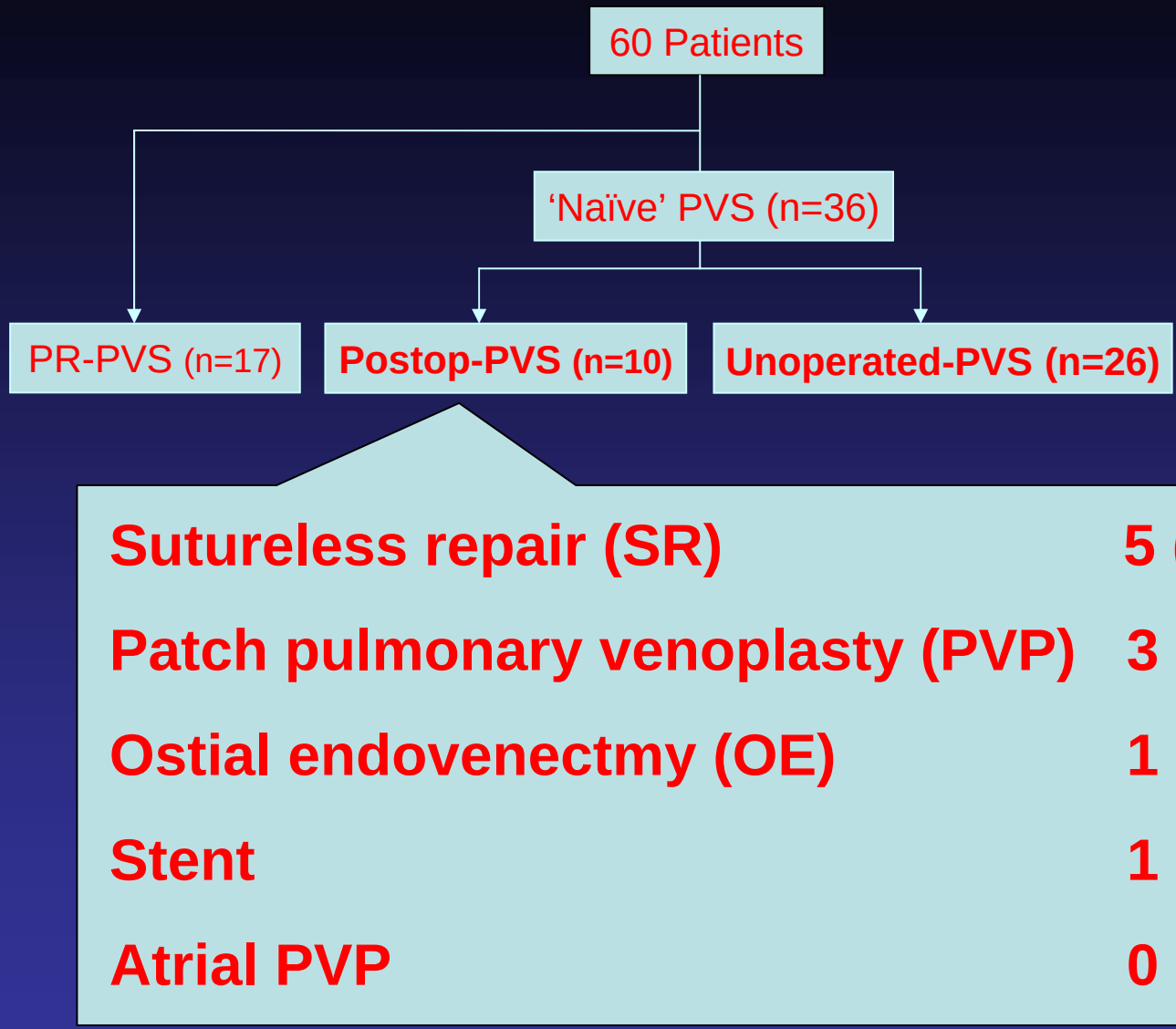
PR-PVS (n=17)

Sutureless repair (SR)	**5 (0)***
Patch pulmonary venoplasty (PVP)	**6 (5)***
Ostial endovenectmy (OE)	**2 (0)***
Stent	**3 (2)***
Atrial PVP	**1 (1)***

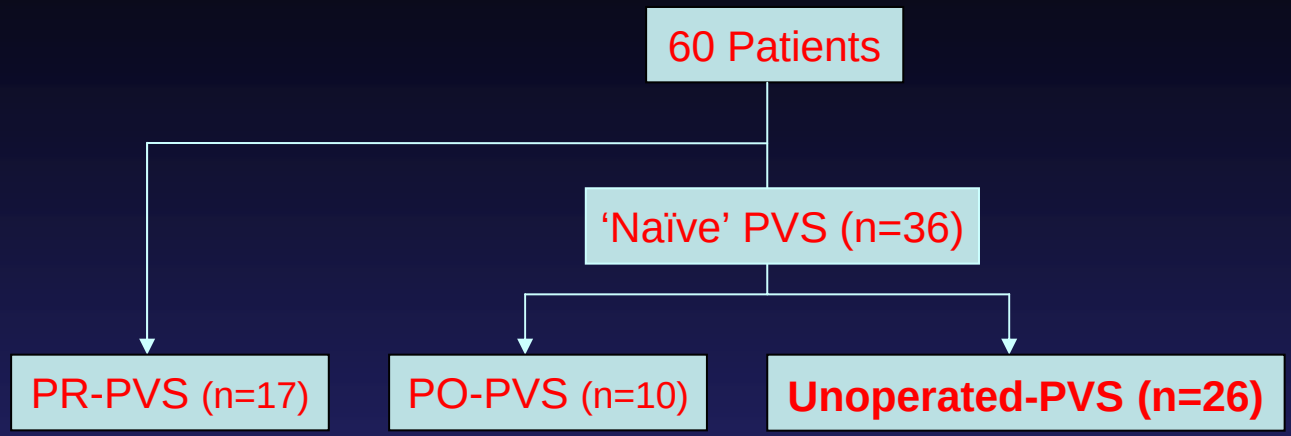
*** Reop or death**


60 Patients
‘Naïve’ PVS (n=36)
PR-PVS (n=17)


60 Patients
‘Naïve’ PVS (n=36)
PR-PVS
Postop-PVS (n=10)
Unoperated-PVS (n=26)

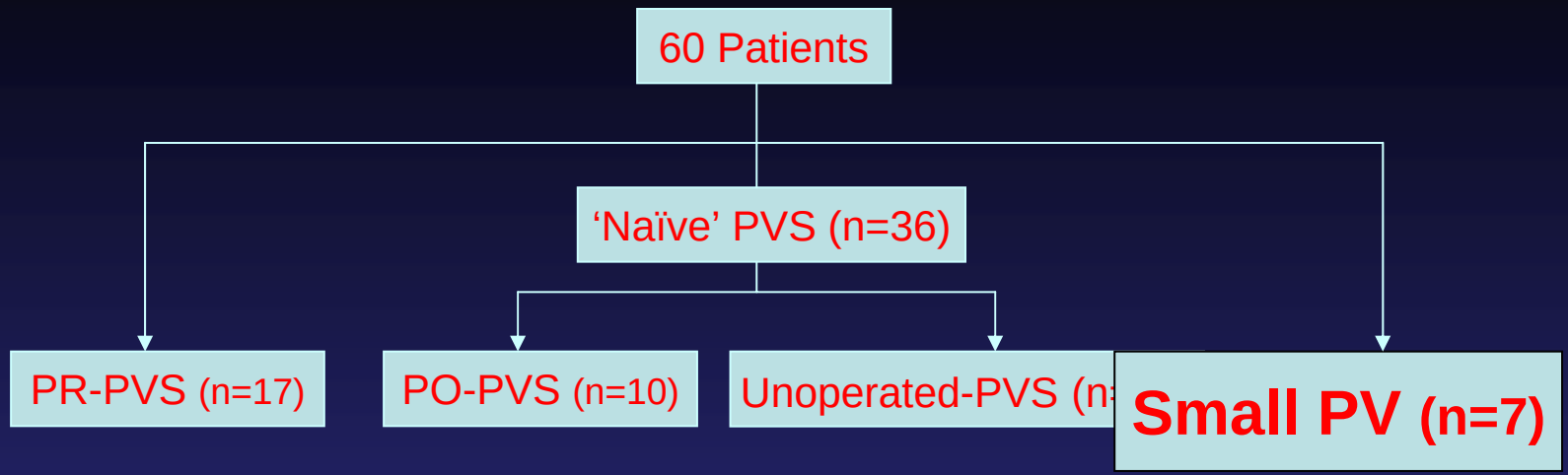


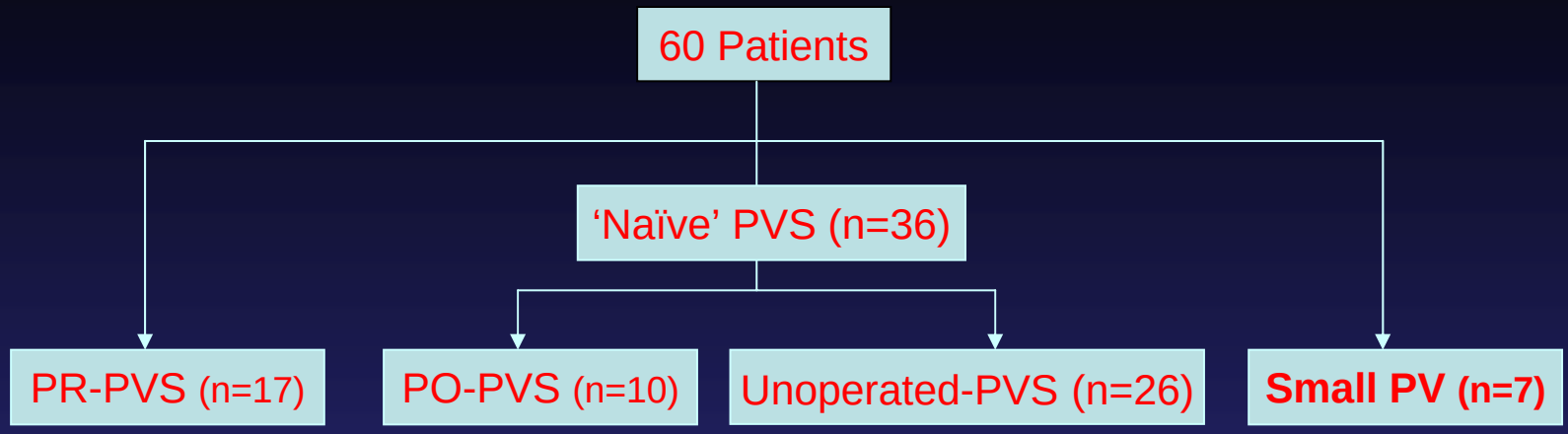
*** Reop or death**



Sutureless repair (SR)	**18 (6)***
Patch pulmonary venoplasty (PVP)	**1 (1)**
Ostial endovenectmy (OE)	**0 (0)**
Stent	**3 (2)**
Atrial PVP	**4 (1)**

*** Reop or death**

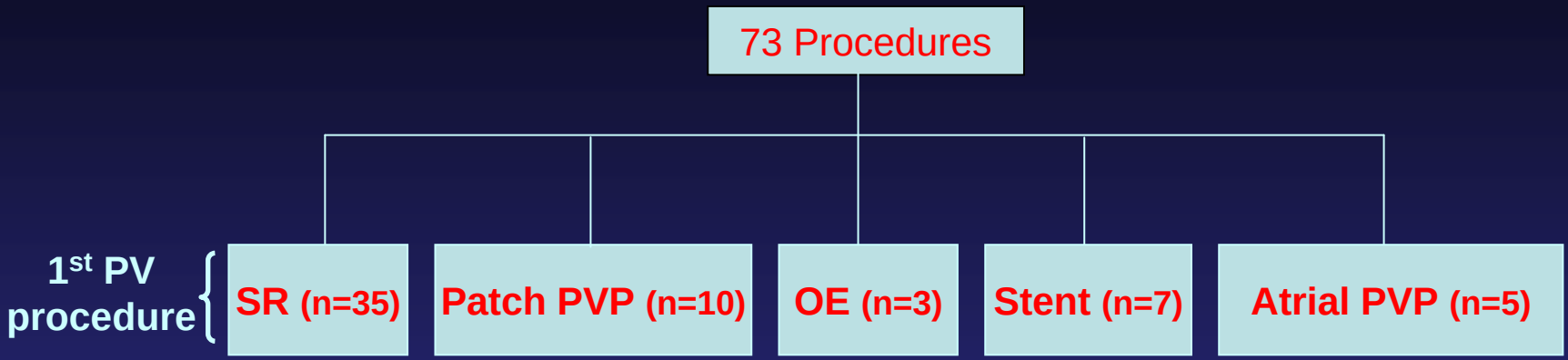

60 Patients
'Naïve' PVS (n=36)
PR-PVS (n=17)
PO-PVS (n=10)
Unoperated-PVS (n=
Small PV (n=7)

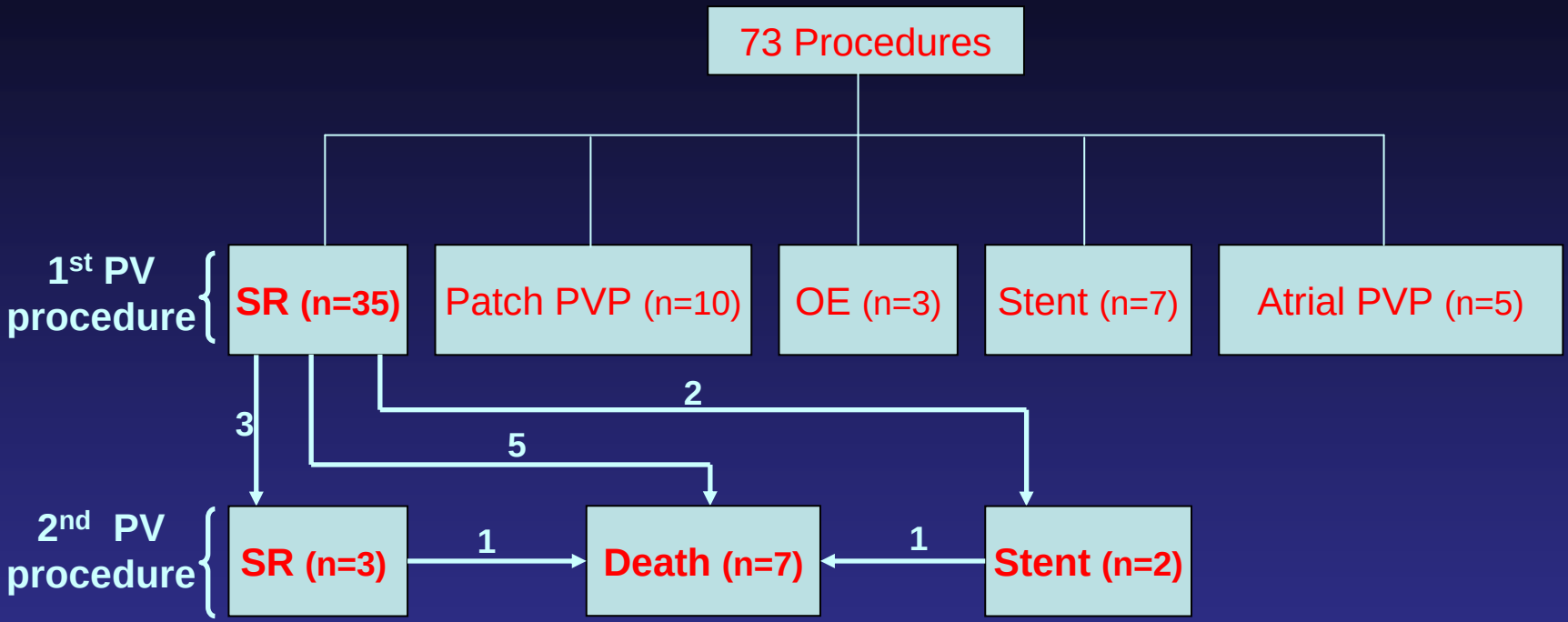


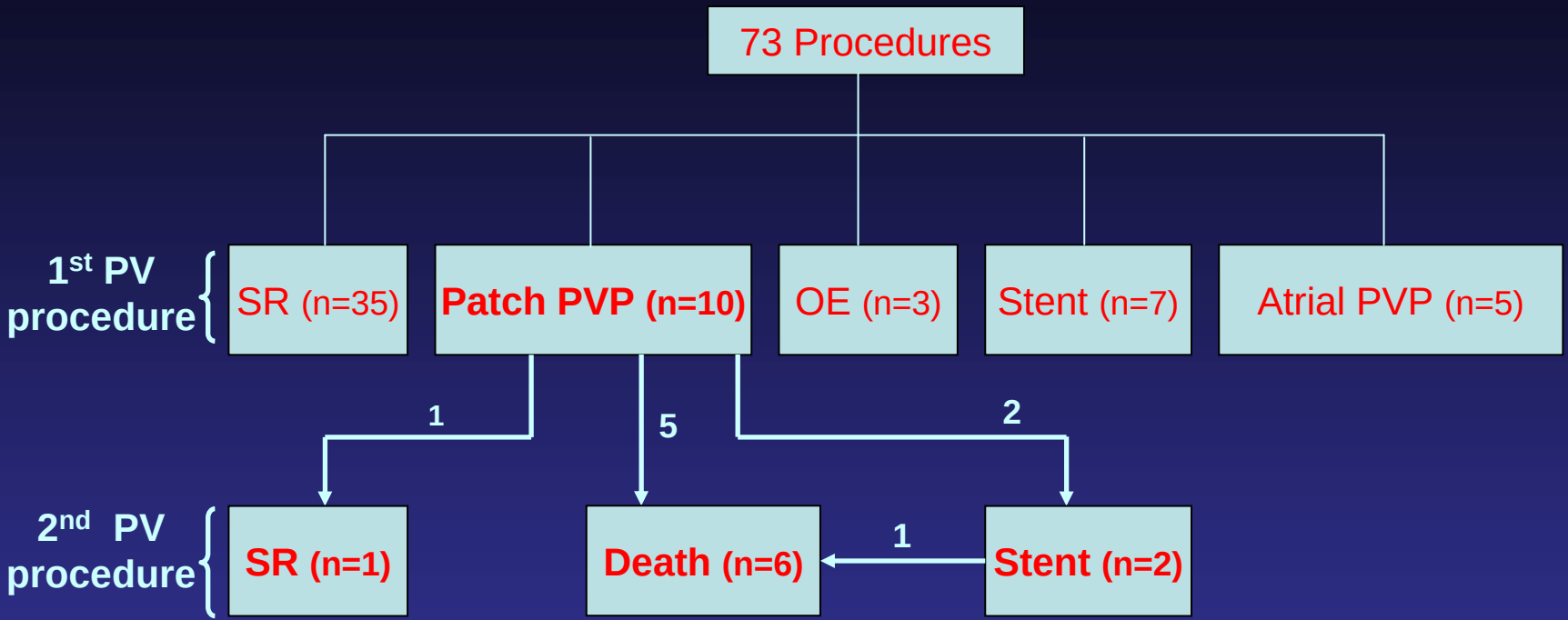
Sutureless repair (SR)	**7 (3)***
Patch pulmonary venoplasty (PVP)	**0 (0)**
Ostial endovenectmy (OE)	**0 (0)**
Stent	**0 (0)**
Atrial PVP	**0 (0)**

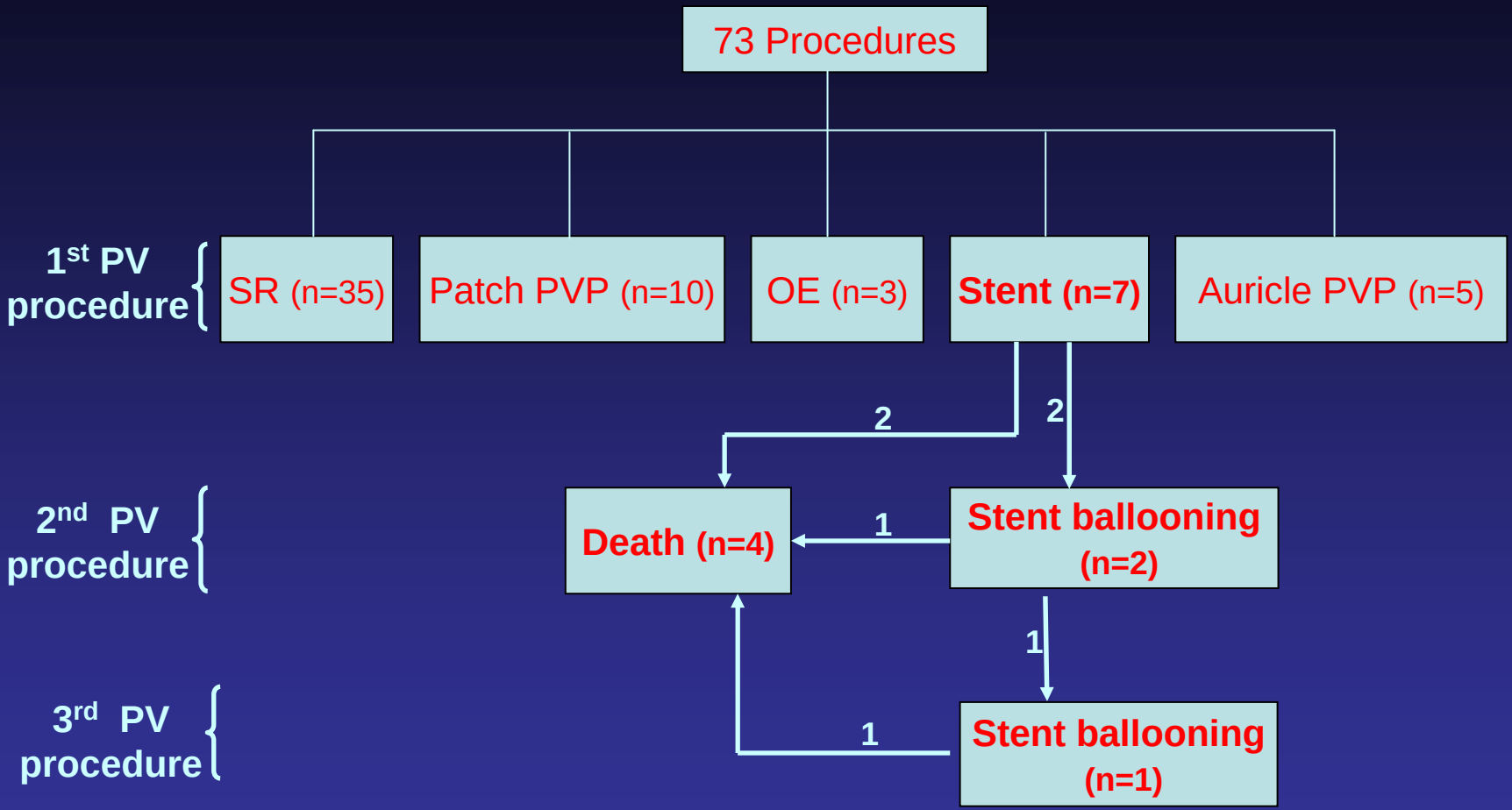
*** Reop or death**

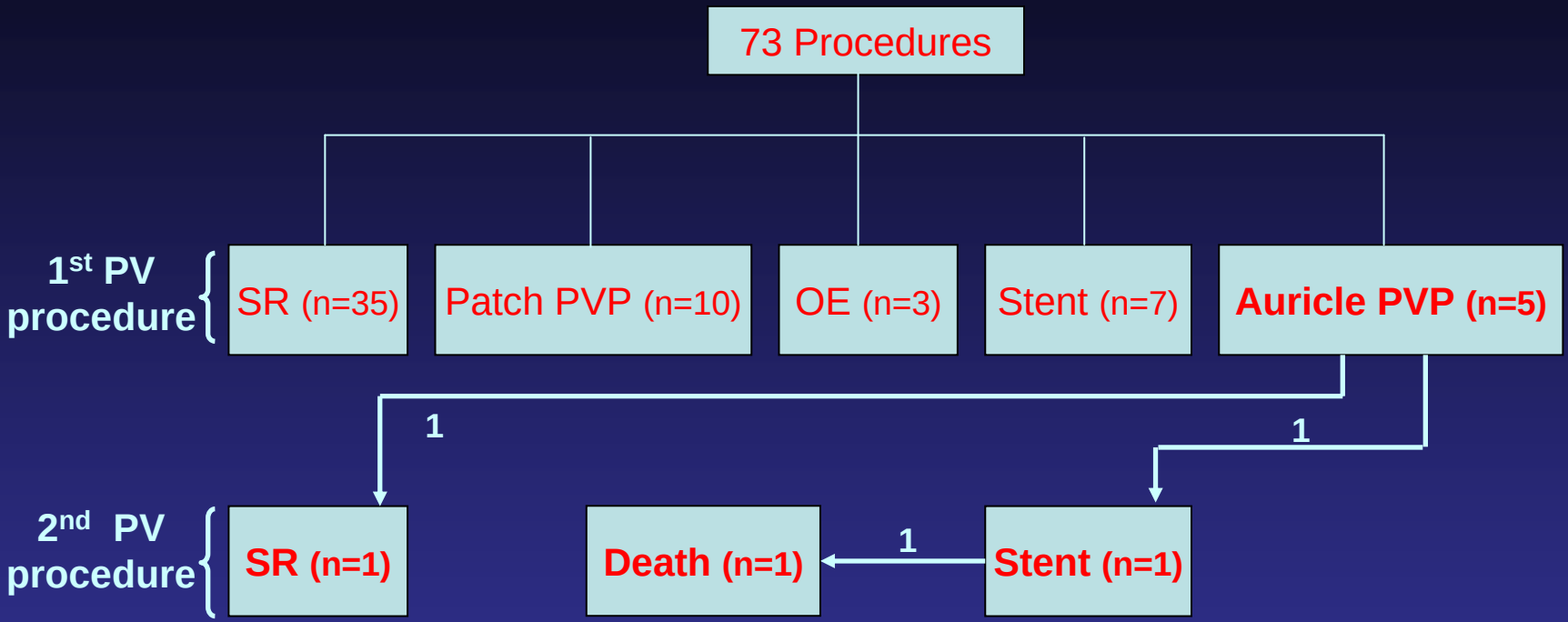
73 Procedures

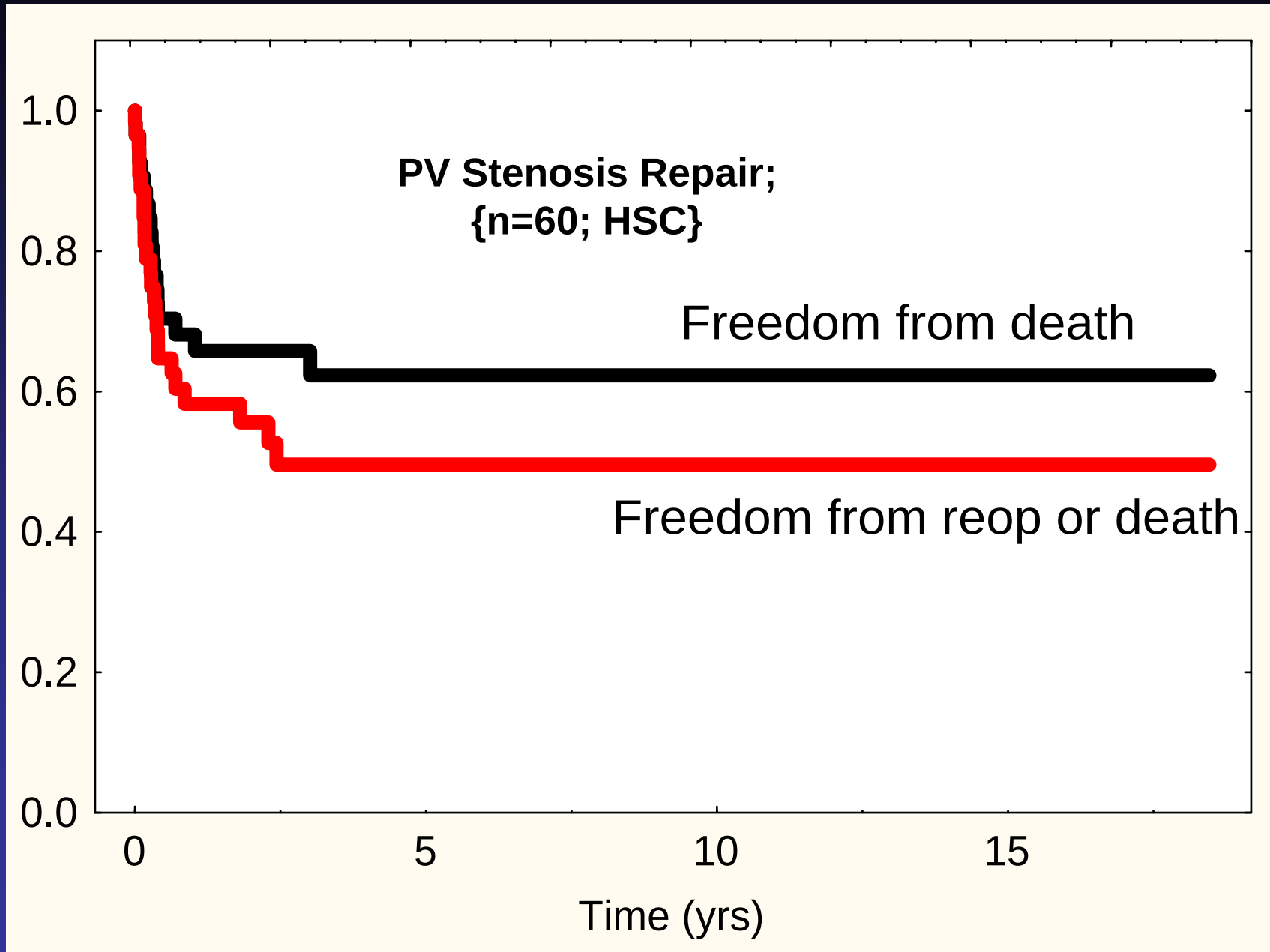

73 Procedures
1st PV procedure
SR (n=35)
Patch PVP (n=10)
OE (n=3)
Stent (n=7)
Atrial PVP (n=5)


73 Procedures
1st PV procedure
SR (n=35)
Patch PVP (n=10)
OE (n=3)
Stent (n=7)
Atrial PVP (n=5)
3
2
5
2nd PV procedure
SR (n=3)
1
Death (n=7)
1
Stent (n=2)


73 Procedures
1st PV procedure
SR (n=35)
Patch PVP (n=10)
OE (n=3)
Stent (n=7)
Atrial PVP (n=5)
1
5
2
2nd PV procedure
SR (n=1)
Death (n=6)
1
Stent (n=2)


73 Procedures
1st PV procedure
SR (n=35)
Patch PVP (n=10)
OE (n=3)
Stent (n=7)
Auricle PVP (n=5)
2
2
2nd PV procedure
Death (n=4)
1
Stent ballooning (n=2)
1
3rd PV procedure
1
Stent ballooning (n=1)


73 Procedures
1st PV procedure
SR (n=35)
Patch PVP (n=10)
OE (n=3)
Stent (n=7)
Auricle PVP (n=5)
1
1
2nd PV procedure
SR (n=1)
Death (n=1)
1
Stent (n=1)


PV Stenosis Repair;
{n=60; HSC}
Freedom from death
Freedom from reop or death
1.0
0.8
0.6
0.4
0.2
0.0
0
5
10
15
Time (yrs)

Statistical Analysis

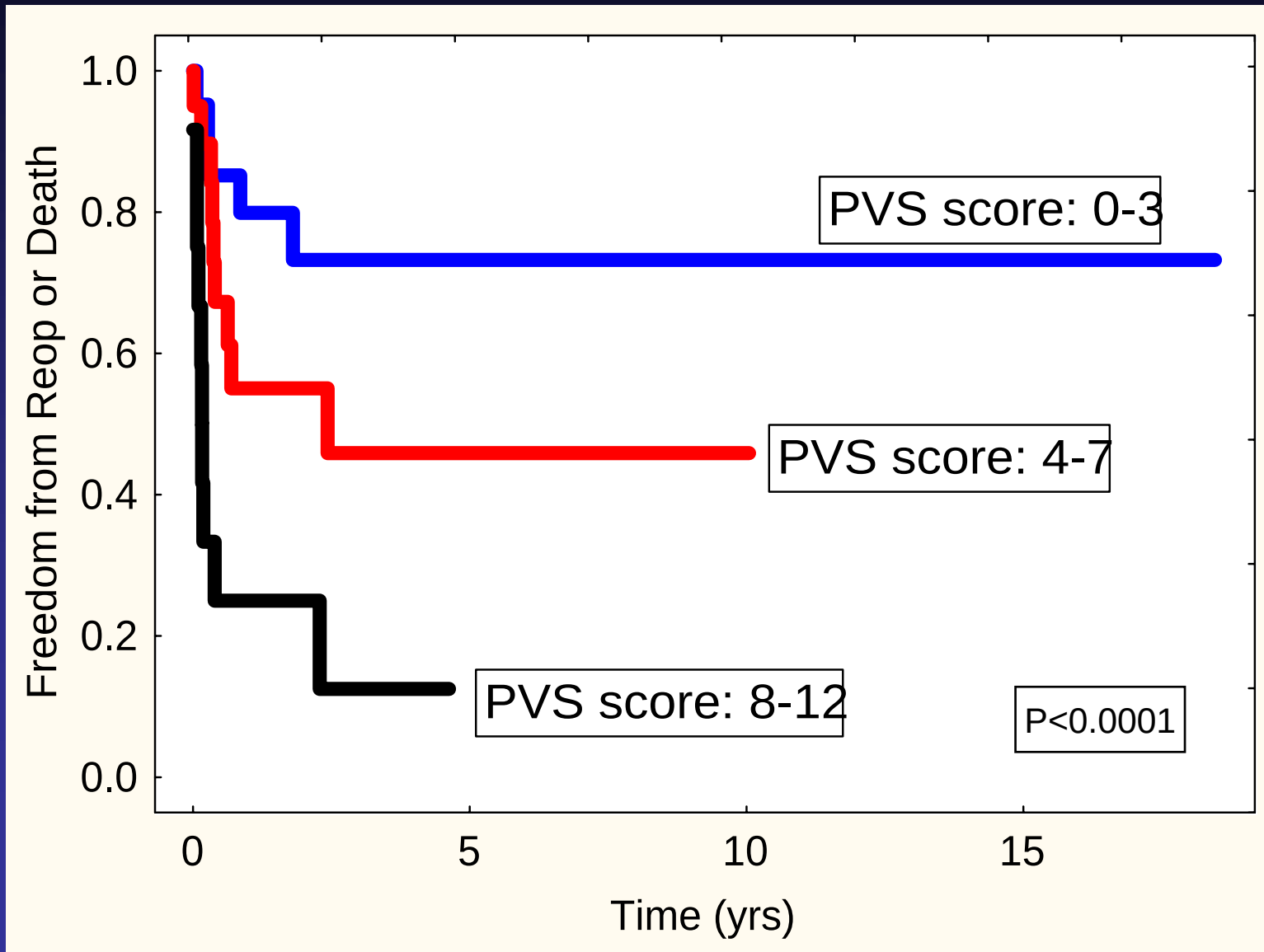
By multivariable analysis

Higher PVS score was associated with increased risk of re-operation or death

HR 12.9 for PVS score from 2-7 p=0.0001

Model validation failed bootstrap analysis

PVS score

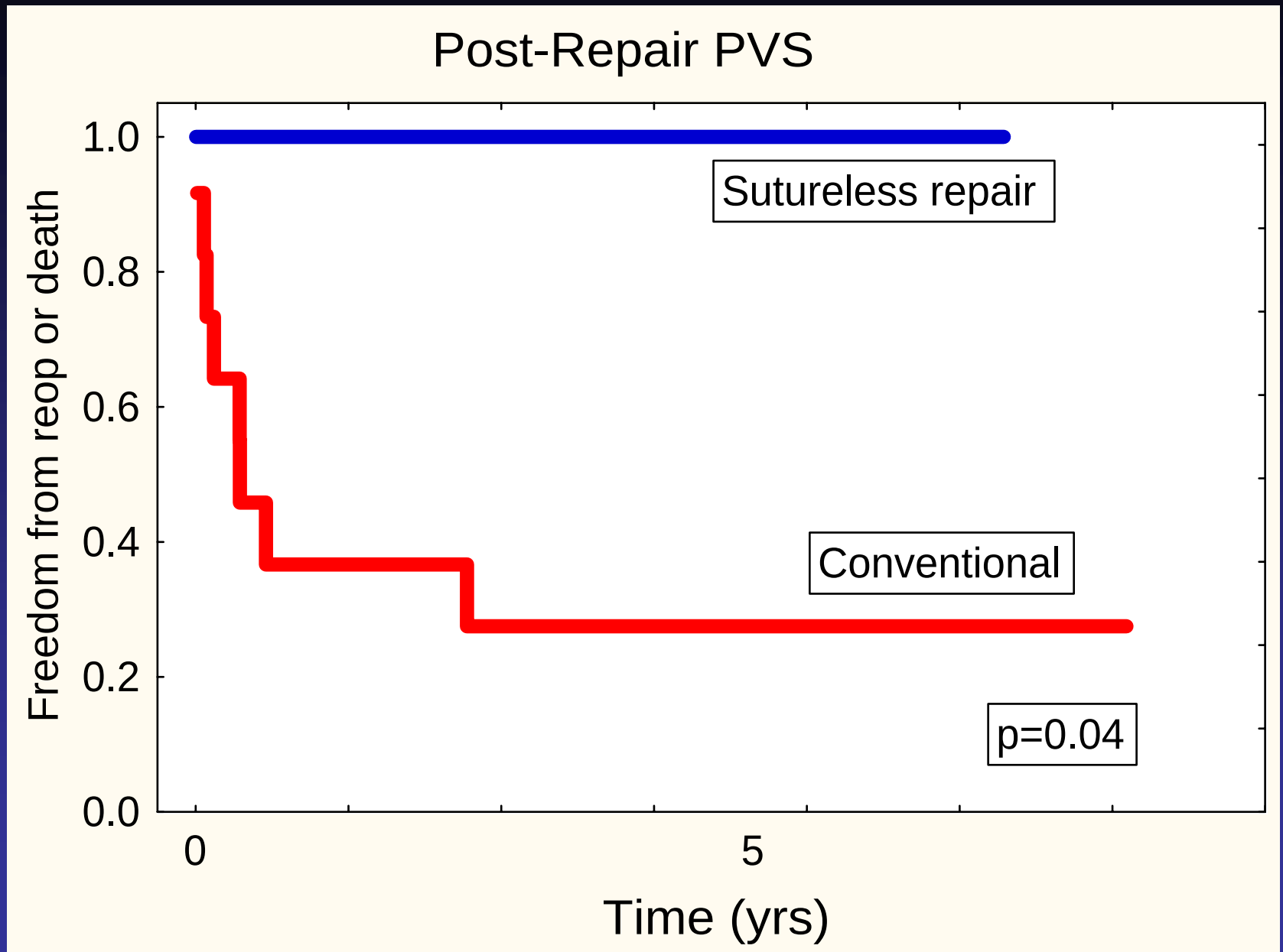

1.0
0.8
0.6
0.4
0.2
0.0
Freedom from Reop or Death
PVS score: 0-3
PVS score: 4-7
PVS score: 8-12
P<0.0001
0
5
10
15
Time (yrs)

Statistical Analysis

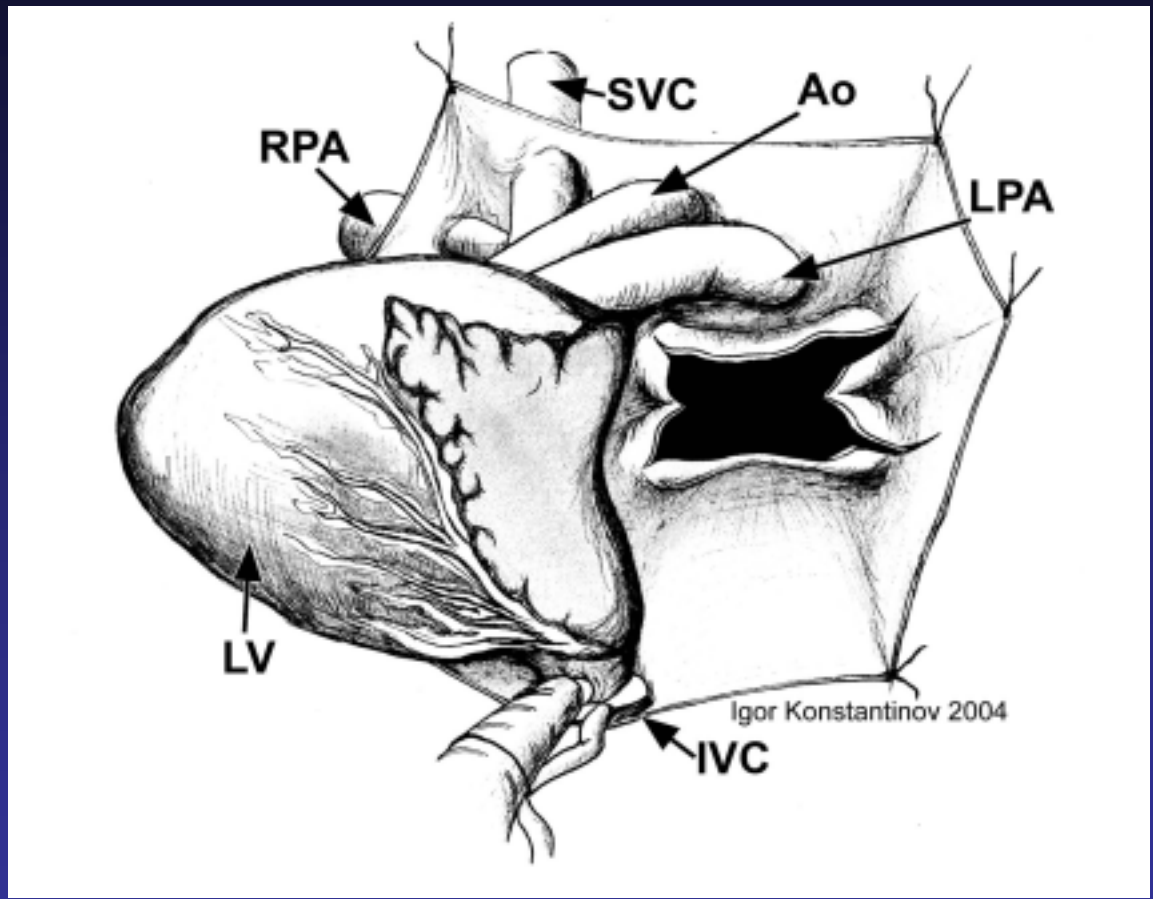
After adjustment for PVS score, the sutureless repair was associated with decreased risk of re-operation or death

HR 0.47 **p=0.12**

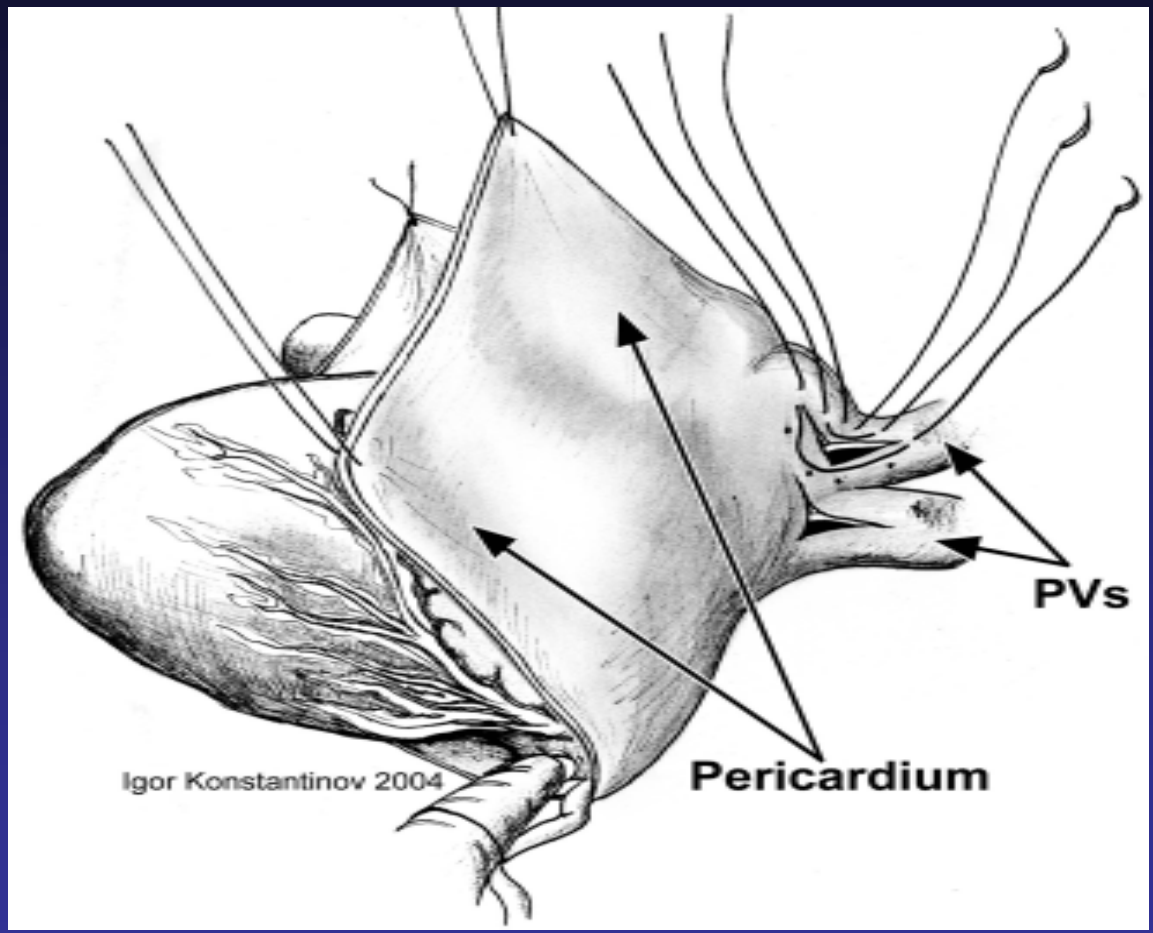
This effect was greatest in the patients with PVS after repair of TAPVD


Post-Repair PVS
Freedom from reop or death
1.0
0.8
0.6
0.4
0.2
0.0
0
5
Time (yrs)
Sutureless repair
Conventional
p=0.04

SR without Retro-cardiac adhesion



SR without Retro-cardiac adhesion



Summary (1)

Sutureless repair (SR) for Post-repair PVS is associated with excellent mid-term results.

PVS score is a significant risk factor for re-operation or death

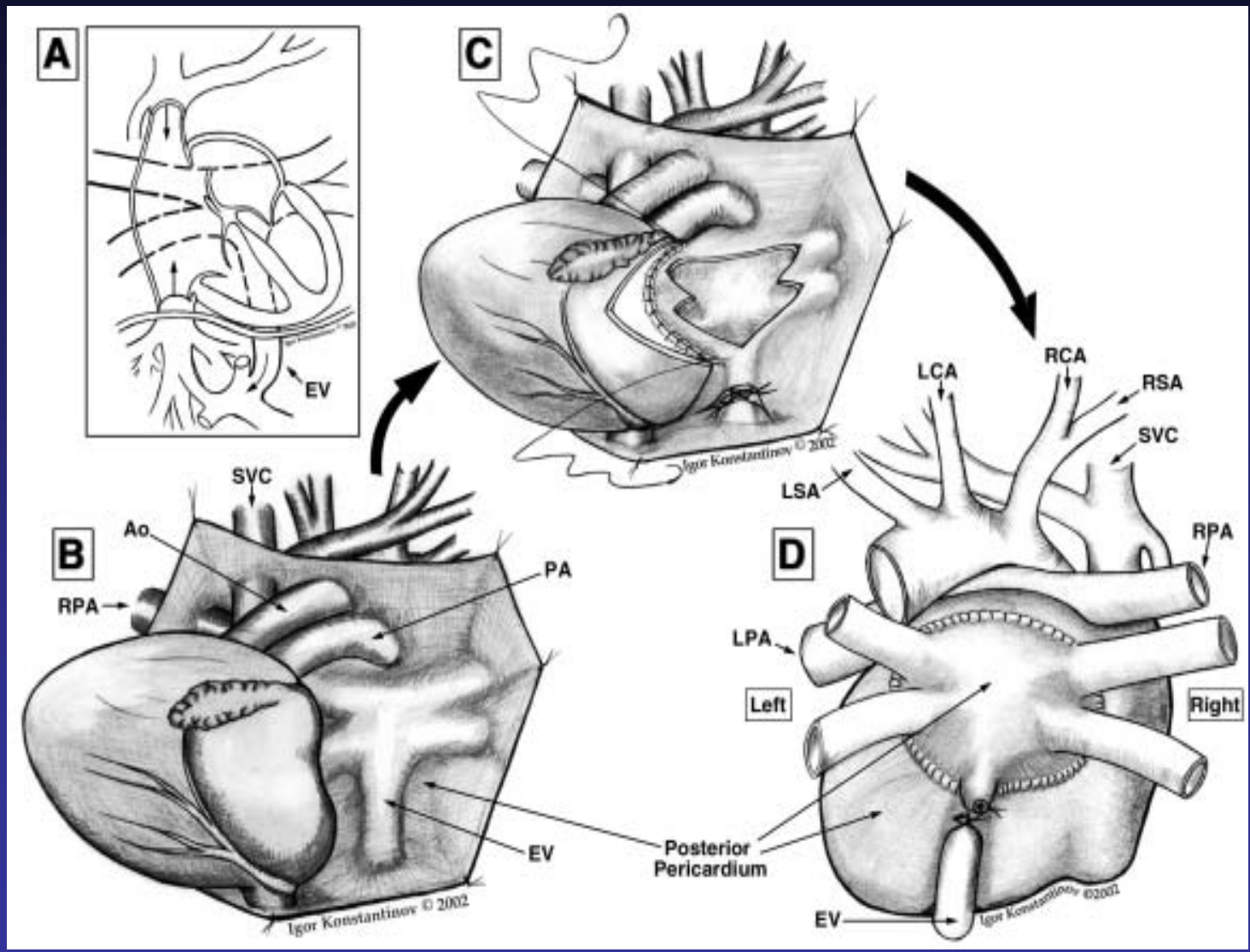
Summary (2)

Sutureless repair without retrocardiac adhesion appears safe with development of a simple hemostatic maneuver

The efficacy of the sutureless repair in patients with indications other than PR-PVS is not well defined due to the small number of patients in the present report

Consequently, a registry-based approach to evaluation would be expedient

SR for TAPVD with small PV/CPVC



Right isomerism with TAPVD

Yun TJ, Osman Al-Radi, Caldarone CA, Coles JG, Williams WG, Smallhorn J, Van Arsdell GS

The Hospital for Sick Children in Toronto

Right atrial isomerism (RAI)

- Failure of lateralization with bilateral right-sidedness
- Centrally squeezed primary PV with abnormal PV drainage
- Dismal prognosis

Management and Outcome of RAI: A 26 year experience
(Hashmi et al, JACC 1998)

- The Hospital for Sick Children in Toronto
- 91 RAI patients between 1970 and 1996
- Cardiac abnormality

  AVSD (81%), FSV (73%), PS (84%)
  TAPVD (87%, 1/3 obstructive)
- No Tx in 24% of patients (with 95% mortality)
- Overall mortality: 69%
- Surgical mortality for TAPVD repair: 95%

Surgical management of TAPVD : Impact of coexisting cardiac anomalies

(Caldarone CA et al, ATT 1998)

- The Hospital for Sick Children in Toronto
- 170 TAPVD pts between 1982 and 1996
- 44 complex TAPVD
- Op. mortality of complex TAPVD: 52%
- Risk factors for Op. mortalities:
  - FSV (P=0.03)
  - Associated complex anomalies (P<0.01)

TAPVD association in RAI

Incidence: 60% - 100%

(Varies according to the definition of TAPVD)

Outcome variables:

1. Draingage site obstruction
2. Individual PVS
3. Presence or absence of TAPVD
4. Types of TAPVD: Cardiac type?

Objective

Premise:

RAI associated with TAPVD is fatal

Question:

Can we modify the outcome by aggressive Tx of TAPVD?

Methods

55 Patients of RAI over 10 years

Patient enrolled: All patients of RAI

Including

1) **Patients without Surgical Tx**
2) **Patients without TAPVD repair**
3) **Patients with TAPVD repair**

Statistical Analysis

Retrospective analysis

Cox Proportional Hazard Survival Model

Observation starting point / end point

: Birth / Re-operation or death

Variables: **Age**

Types of TAPVD

Obstructive vs. non-obstructive

FSV vs. BVR candidate

PBF: ↑ or ↓ or ↔

Types of intervention (SR vs. Others)

Model validation with bootstrap analysis

Withdrawal (n=11)

Decision making of withdrawal

by parents: 7 / 11

by co-existing medical problem: 4/11

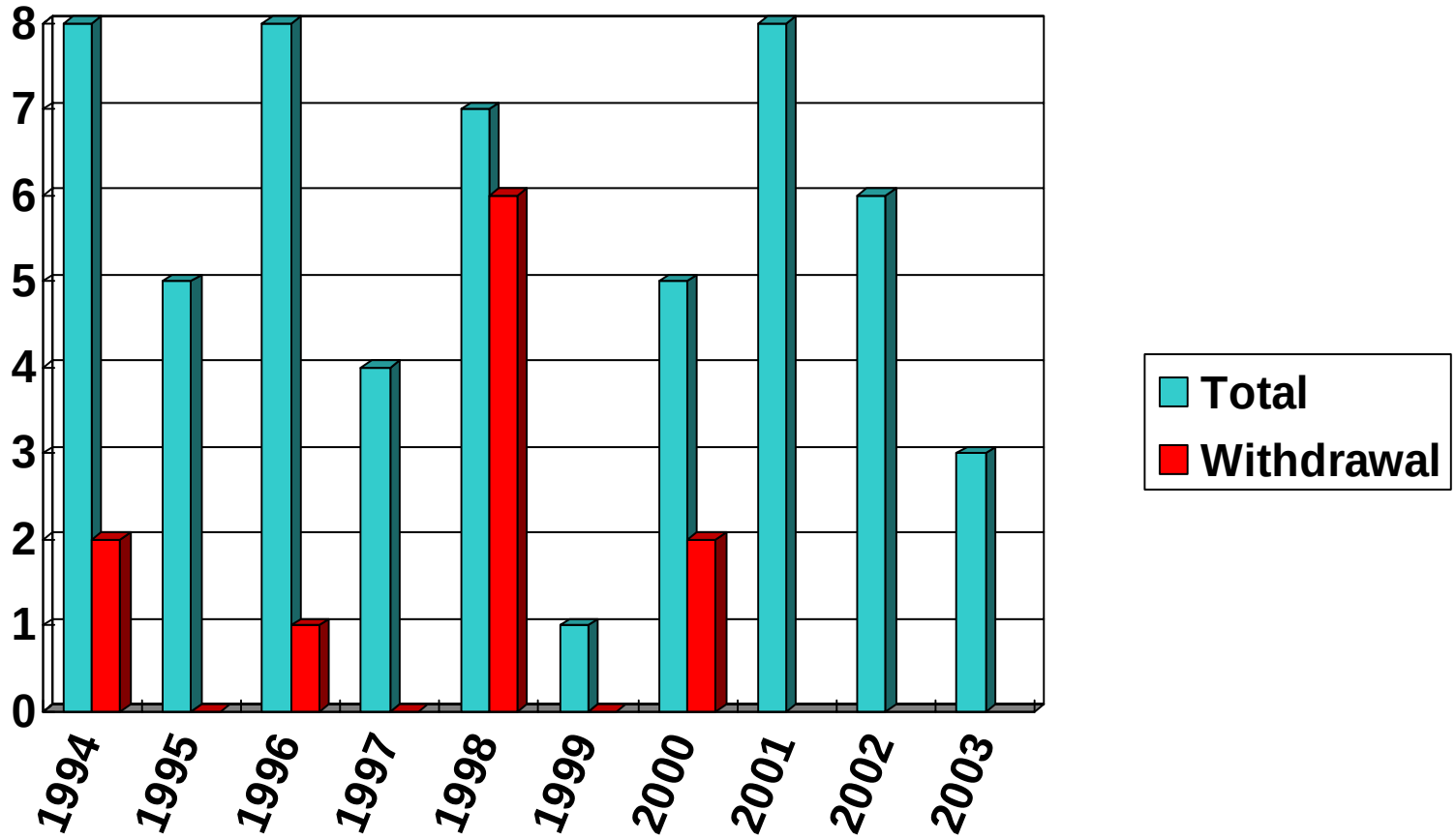
- Chromosomal anomaly (n=1)
- Delayed Dx with MOF (n=1)
- Complex obstructive TAPVD (n=2)

High withdrawal rate by specific cardiologist

	Cardiologist A	Others
Tx	11	33
No Tx	7	4
Total	18	37

(P=0.000326, OR=12.2)

Annual changes in withdrawal



Types of TAPVD (n=55)

Group	No TAPVD	SupraC	Cardiac	InfraC	Mixed
No Tx	0	5(2)	1(0)	1(1)	4(3)
TAPVD repair (-)	4(0)	3(1)	3(0)	0	3(0)
TAPVD repair (+)	0	19(7)	5(4)	5(3)	2(1)

TAPVD, Total anomalous pulmonary venous drainage; Tx, Treatment; ( ), Draining site obstruction

Operative procedures

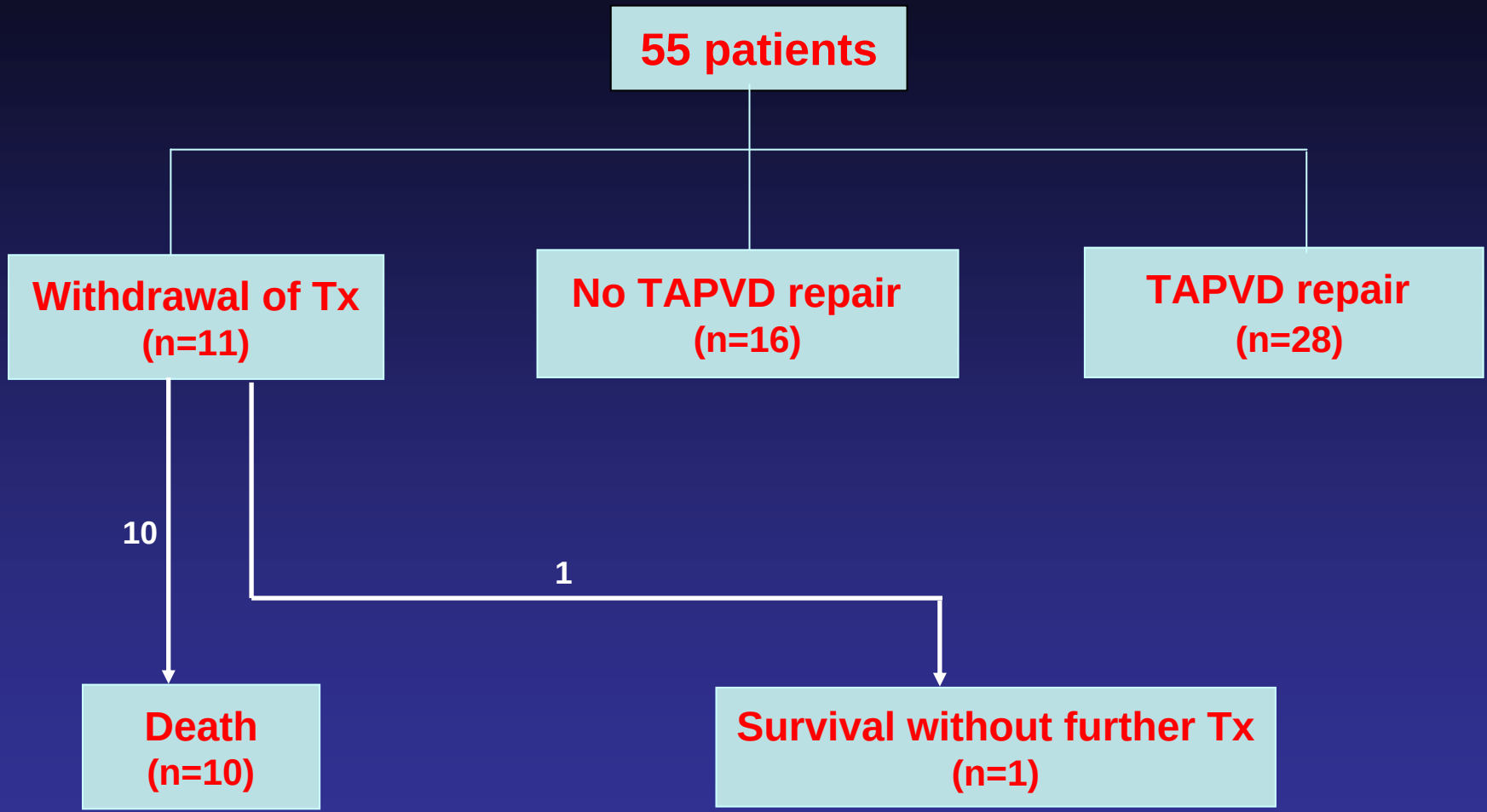
	Without TAPVD repair	With TAPVD repair
Systemic-pulmonary shunt	7	7
PA banding	2	4
BCPS	1	9
Fontan operation	1	3
Cardiac Transplantation	2	2
PA angioplasty	0	7
Isolated TAPVD repair	0	4
Norwood operation	2	0
Miscellaneous*	0	3

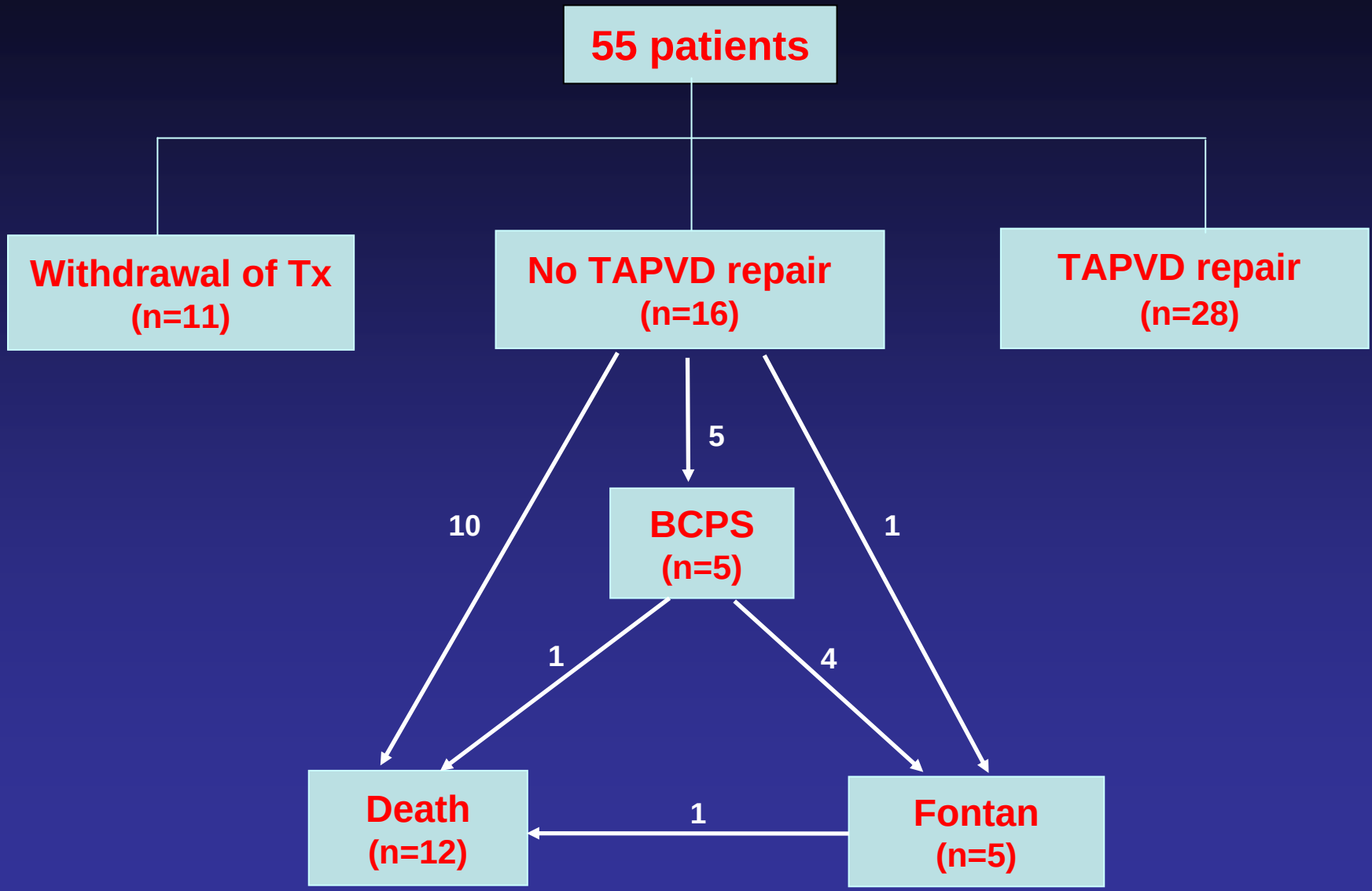
Ventricular Morphology and Surgical Strategies

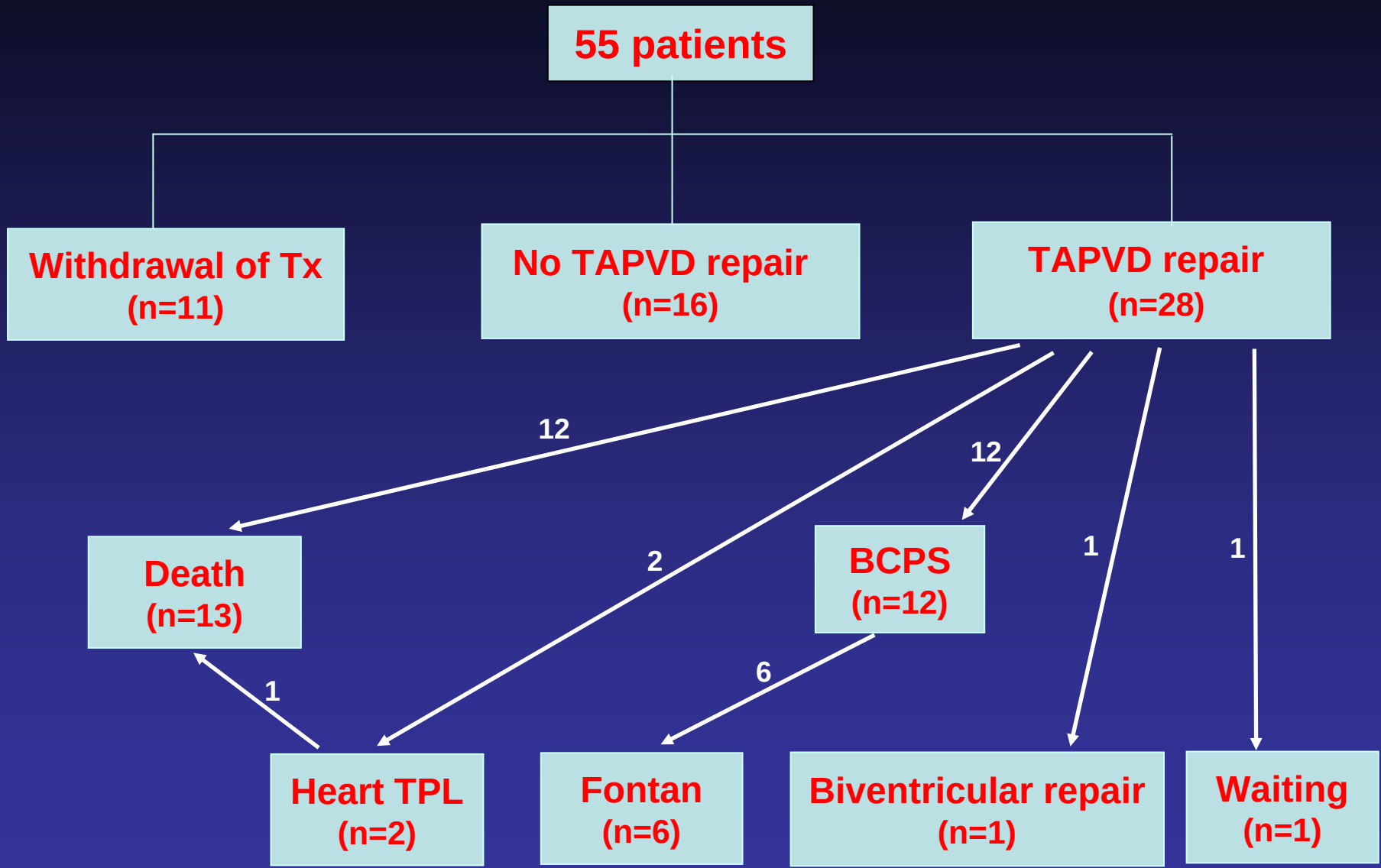
	No Tx	BVR	OVR	HTx	Undetermined*
Balanced ventricles (n=20)	2	2	5	1	10
Unbalanced ventricles (n=35)	9	0	16	1	9

Tx, treatment; BVR, Biventricular repair; OVR, One ventricle repair; HTx, Heart Transplantation

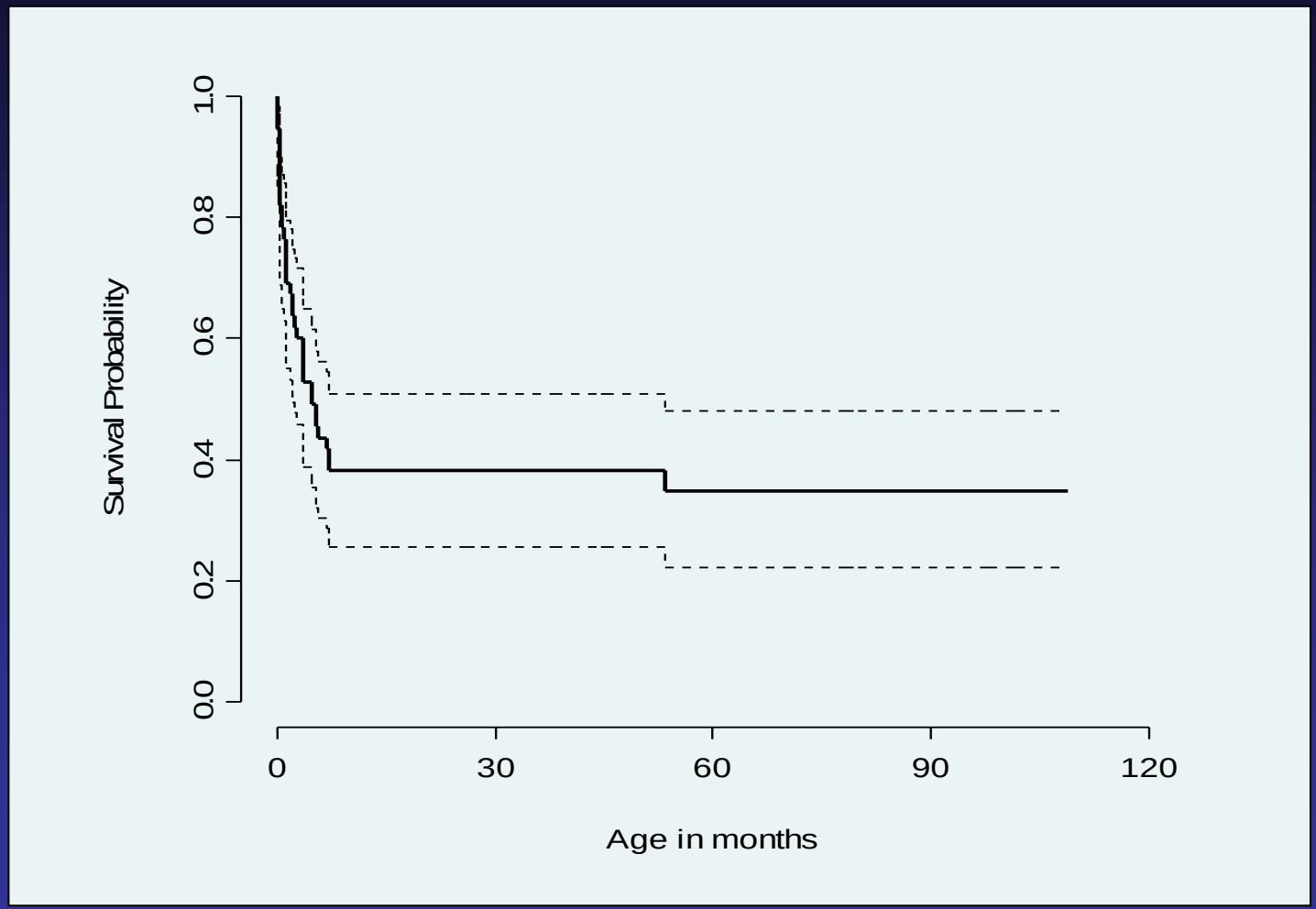
* Undetermined: operative death or treatment withdrawal before determination of surgical strategy


55 patients
Withdrawal of Tx
(n=11)
No TAPVD repair
(n=16)
TAPVD repair
(n=28)
10
1
Death
(n=10)
Survival without further Tx
(n=1)

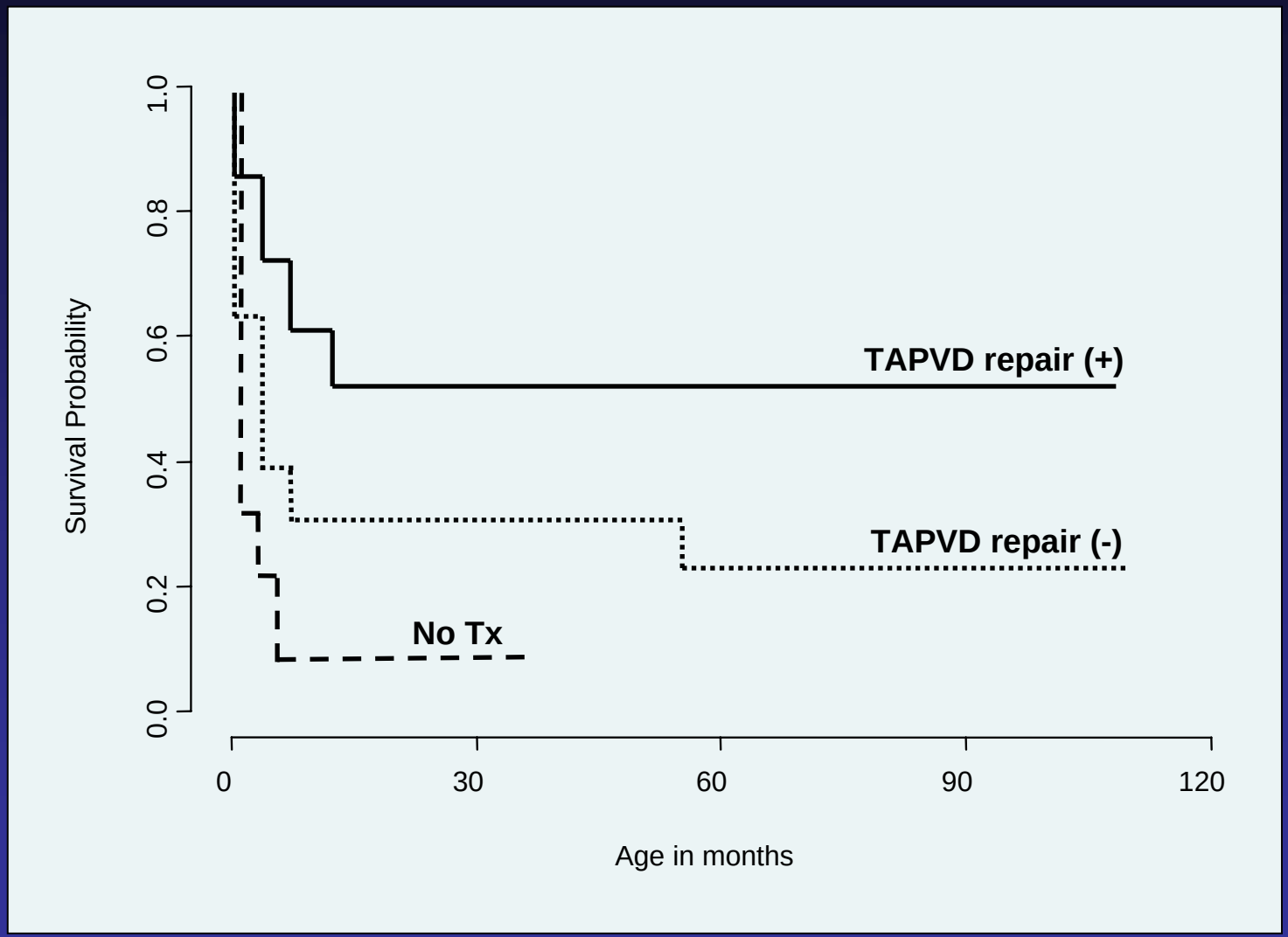

55 patients
Withdrawal of Tx
(n=11)
No TAPVD repair
(n=16)
TAPVD repair
(n=28)
5
10
1
BCPS
(n=5)
1
4
Death
(n=12)
1
Fontan
(n=5)


55 patients
Withdrawal of Tx (n=11)
No TAPVD repair (n=16)
TAPVD repair (n=28)
12
12
2
1
6
1
1
Death (n=13)
BCPS (n=12)
Heart TPL (n=2)
Fontan (n=6)
Biventricular repair (n=1)
Waiting (n=1)

Survival after birth (Entire cohort, n=55)



Survival by groups



Summary

- Withdrawal from treatment has poor results with 95% mortality within a year
- Risk factors for death in TAPVD repair group
  1) Obstructive TAPVR
  2) Mixed and infracardiac type TAPVR
- Adjusted with these risk factors, primary sutureless repair for TAPVD provide survival benefit (HR:0.43)

Conclusions

- Most patients with PVS are Surgical candidates
- Sutureless repair for PR-PVS shows excellent long term outcome
- Surgical Tx of Congenital PVS is still challenging, and Sutureless repair might be an option
- Primary Sutureless repair for TAPVD with small PV needs to be defined
- Registry based approach for PVS might be expedient to come up with a conclusion