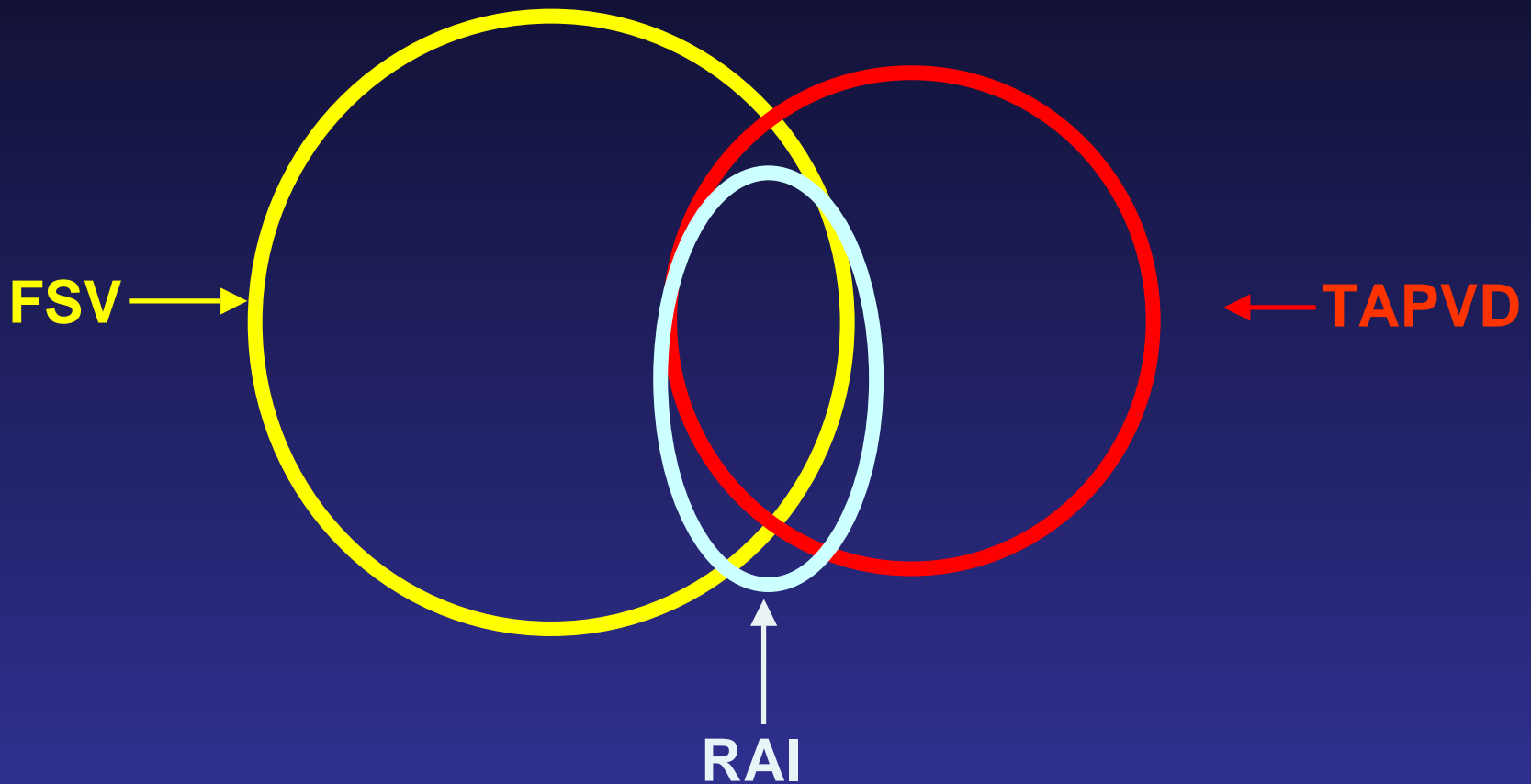


**Functional SV with TAPVD:
Contemporary Management of Right Atrial
Isomerism**

Yun TJ, Van Arsdell GS

*Asan Medical Center
The Hospital for Sick Children in Toronto*

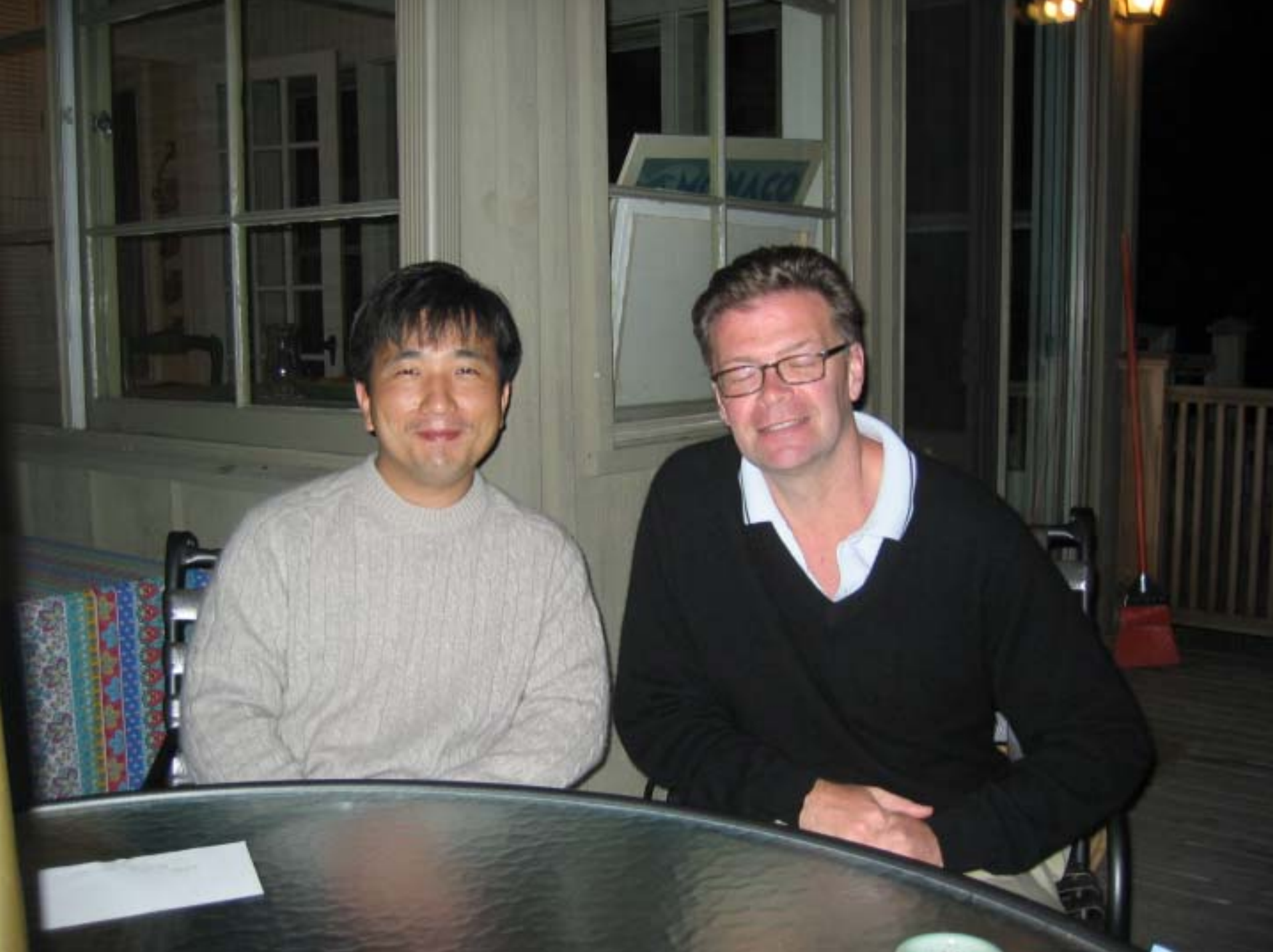
Functional SV, TAPVD and RAI



**Functional SV with TAPVD and
management of postoperative PVS**

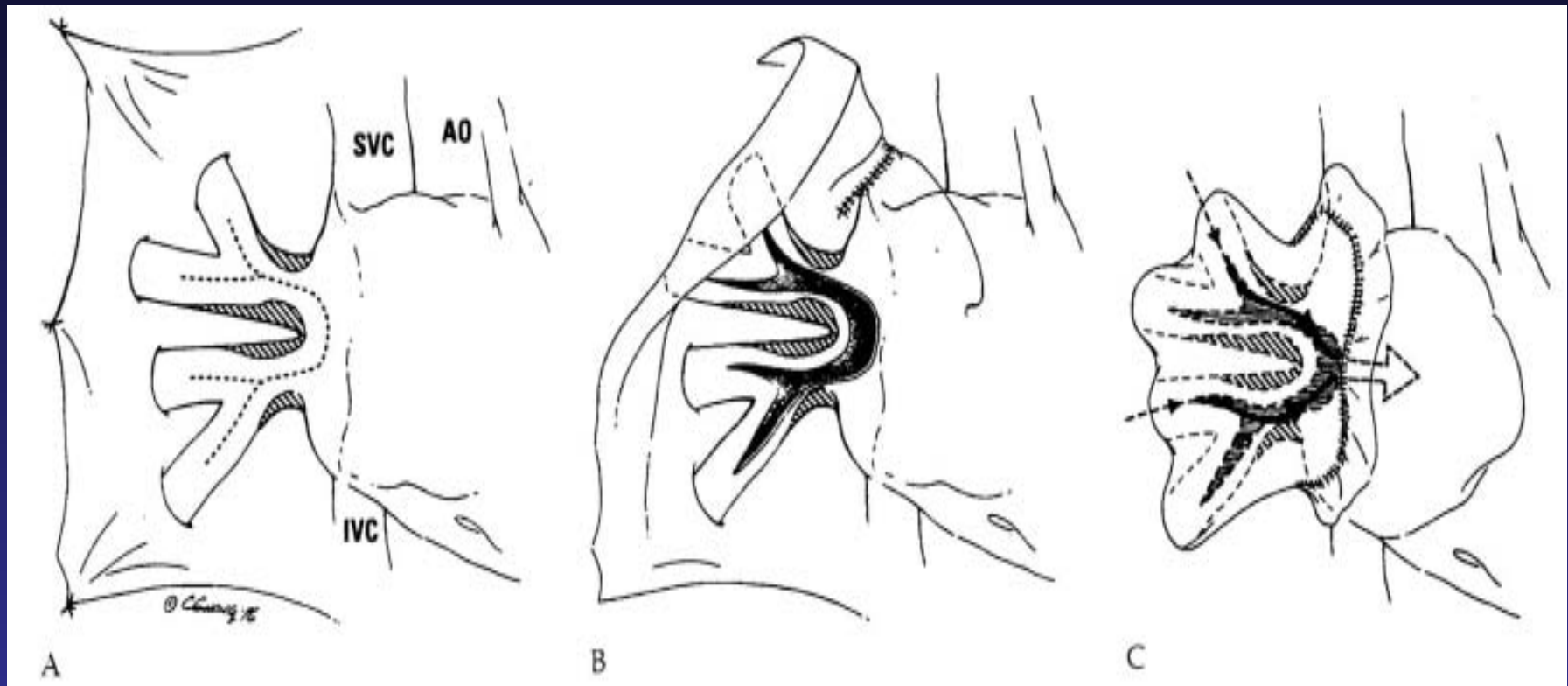


**Contemporary management of
Right atrial isomerism**



A Sutureless technique for PVS

(Najm HK, Caldarone CA, Smallhorn J, Coles JG. JTCS 1998;115(2):468-70)



Surgical management of TAPVD

: Impact of coexisting cardiac anomalies

(Caldarone et al, ATT 1998)

- **The Hospital for Sick Children**
- **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)**



Conventional and sutureless techniques for management of the pulmonary veins: Evolution of indications from postrepair pulmonary vein stenosis to primary pulmonary vein anomalies

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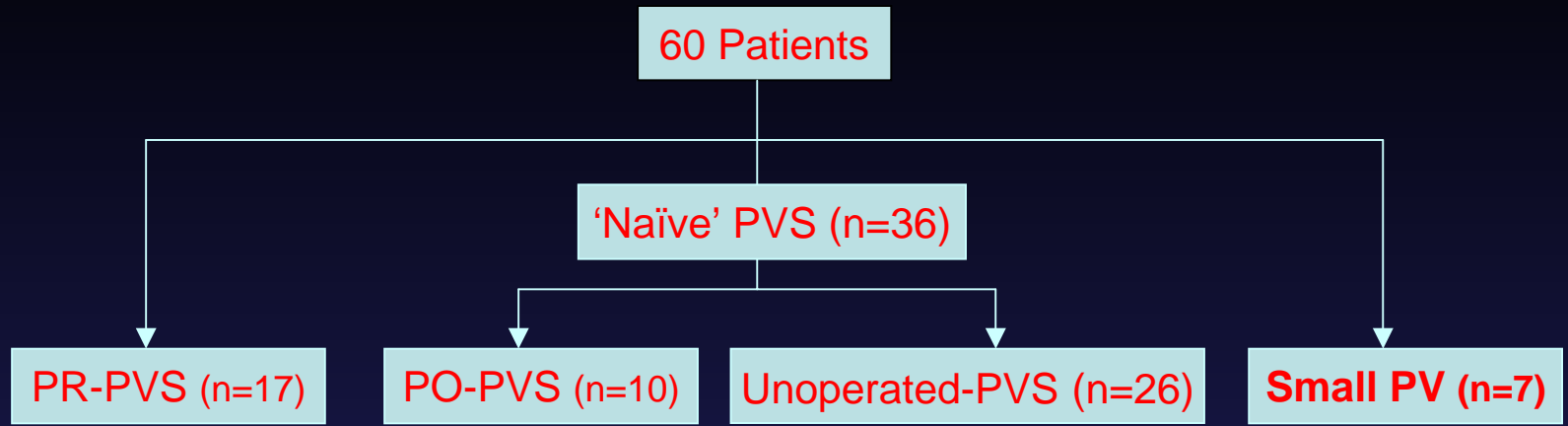
doi:10.1016/j.jtcvs.2004.08.043

Objective: We have previously reported a limited but favorable experience with a novel sutureless technique for surgical management of postoperative pulmonary vein stenosis occurring after repair of total anomalous pulmonary venous drainage. Because this technique requires integrity of the retrocardiac space for hemostasis, extension of the technique to the primary repair of pulmonary vein anomalies requires evaluation. This analysis reviews our experience with the sutureless technique in patients with postrepair pulmonary vein stenosis, as well as our extension of the technique into primary repair of pulmonary vein anomalies.

Methods: Retrospective univariable-multivariable analysis of all pulmonary vein stenosis procedures and sutureless pulmonary vein procedures over a 20-year period was performed. Cox proportional hazards modeling was used to identify variables associated with freedom from reoperation or death.

Results: Sixty patients underwent 73 procedures, with pulmonary vein stenosis present in 65 procedures. The sutureless technique was used in 40 procedures. Freedom from reoperation or death at 5 years after the initial procedure was 49%. Unadjusted freedom from reoperation or death was greater with the sutureless technique for patients with postrepair pulmonary vein stenosis ($P = .04$). By using multivariable analysis, a higher pulmonary vein stenosis score was associated with greater risk of reoperation or death. After adjustment, the sutureless repair was associated with a nonsignificant trend toward greater freedom from reoperation or death ($P = .12$). Despite the absence of retrocardiac adhesions, operative mortality was not increased with the sutureless technique ($P = .64$). Techniques to control bleeding (intrapleural hilar reapproximation) and improve exposure (inferior vena cava division) were identified.

Conclusion: The sutureless technique for postrepair pulmonary vein stenosis is associated with encouraging midterm results. Extension of the indications for the technique to primary repair appears safe with the development of simple intraoperative maneuvers.



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

*** Reop or death**

Management and Outcome of RAI:

A 26 year experience

(Hashmi et al, JACC 1998)

- **The Hospital for Sick Children**
- **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%**

Patient enrollment

Considerations for pt. enrollment

1. Do we include patients without Tx ?
2. Do we include patients without TAPVD?
3. Definition of cardiac type TAPVD ?
4. Do we include patients without PV operations ?
5. Starting point of observation for patients without Tx or TAPVD operation?

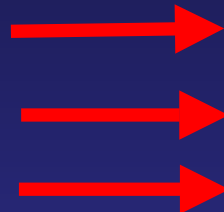
Evolution of Therapeutic Strategies

: by Dr. Glen S. Van Arsdell

2000-2001



Treatment withdrawal
No repair of TAPVD
Direct repair of TAPVD
No ECMO / VAD
Medical Tx of arrhythmia



No Treatment withdrawal
SR for TAPVD repair
ECMO / VAD
Surgical Tx of arrhythmia





WTSA

Annual Meeting Information



31ST ANNUAL MEETING

Victoria, British Columbia
Fairmont Empress Hotel

June 22-25, 2005



Western Thoracic Surgical Association

www.westernthoracic.org

P19. Contemporary Management of Right Atrial Isomerism: Impact of Evolving Therapeutic Strategies

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BACKGROUND: Infants with right atrial isomerism (RAI) have poor outcomes due to complex combination of cardiac anomalies. Aggressive management of total anomalous pulmonary venous drainage (TAPVD) may have a positive impact on the prognosis.

METHODS: Outcomes of all children with RAI from 1994 to present were retrospectively reviewed. Management of TAPVD evolved from no repair or conventional surgical technique to primary sutureless repair (SR) upon initial palliation. Cox survival models were used to identify variables associated with reduced survival.

RESULTS: There were 55 children (34 males). Median age at initial visit was 2 days. Withdrawal of treatment occurred in 11 (11/55, 20%) during an interval of institutional bias towards no treatment. Thirteen patients (13/55, 24%) had palliations without TAPVD repair - 3 survived (3/13, 23%). Thirty-one patients (31/55, 56%) had operations that included TAPVD repair, of whom 13 (13/31, 42%) underwent primary SR for TAPVD. Sixteen patients survived (16/31, 52%), and their current status 1 to 10 yrs (median: 5.8 yrs) after repair is post-Fontan (7/16, 44%), post-bidirectional Glenn (6/16, 38%), and others (3, 20%). In patients who underwent TAPVD repair (n=31), two risk factors of decreased survival were identified: drainage site obstruction, infracardiac or mixed type TAPVD. After adjustment, SR was associated with improved survival, HR of 0.43, but this beneficial effect failed to reach statistical significance (p= 0.19).

CONCLUSIONS: Although mortality is still high, aggressive TAPVD repair for RAI has resulted in improved survival. The role of primary SR for TAPVD associated with RAI has remained to be defined.

The Journal of Thoracic and Cardiovascular Surgery

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Thank you for submitting your manuscript, "WTSA 2005 #P19
**Contemporary Management of Right Atrial Isomerism: Impact of Evolving
Therapeutic Strategies**," to The Journal of Thoracic and Cardiovascular
Surgery. Three highly knowledgeable individuals and I have read your
manuscript and find that it is **potentially acceptable**. However, the
manuscript will require additional review following revision. The comments
of the reviewers are included below.

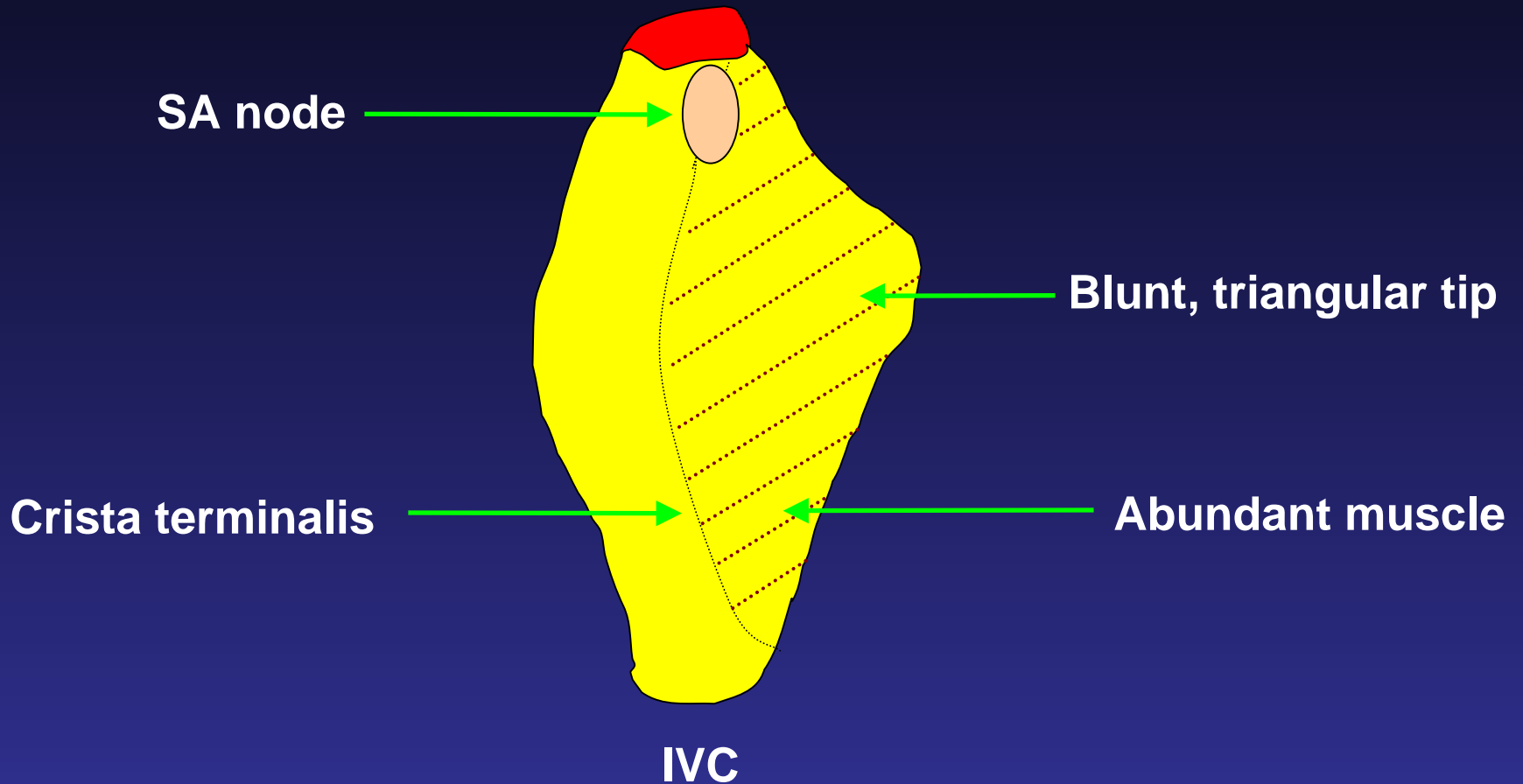
Reviewer #1: This is a very interesting paper evaluating outcomes for
infants with right atrial isomerism. Many of these children have
functional single ventricle and total anomalous pulmonary venous drainage.
Previous studies from several institutions have suggested that the outcome
for these patients is very poor.



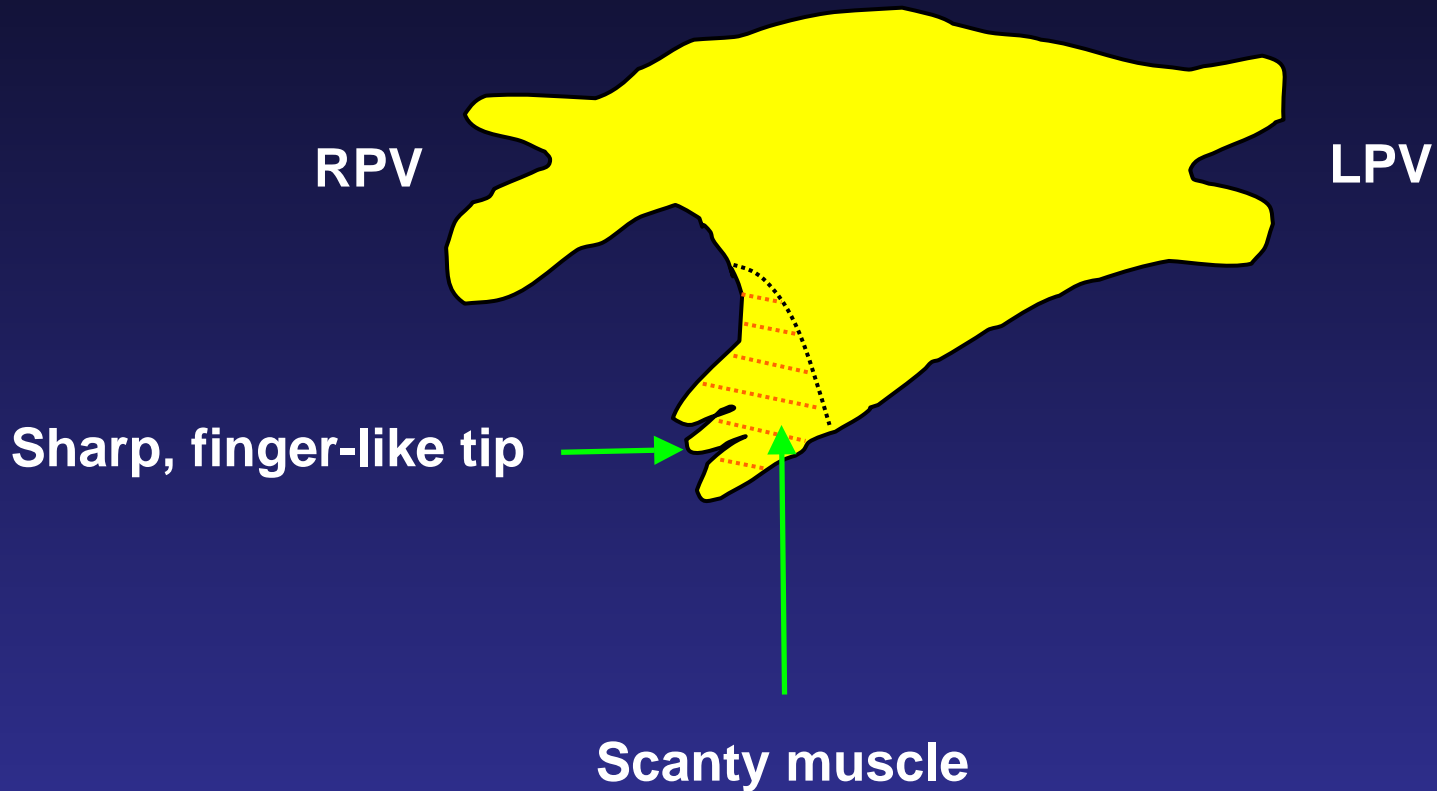
Right atrial isomerism (RAI)

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

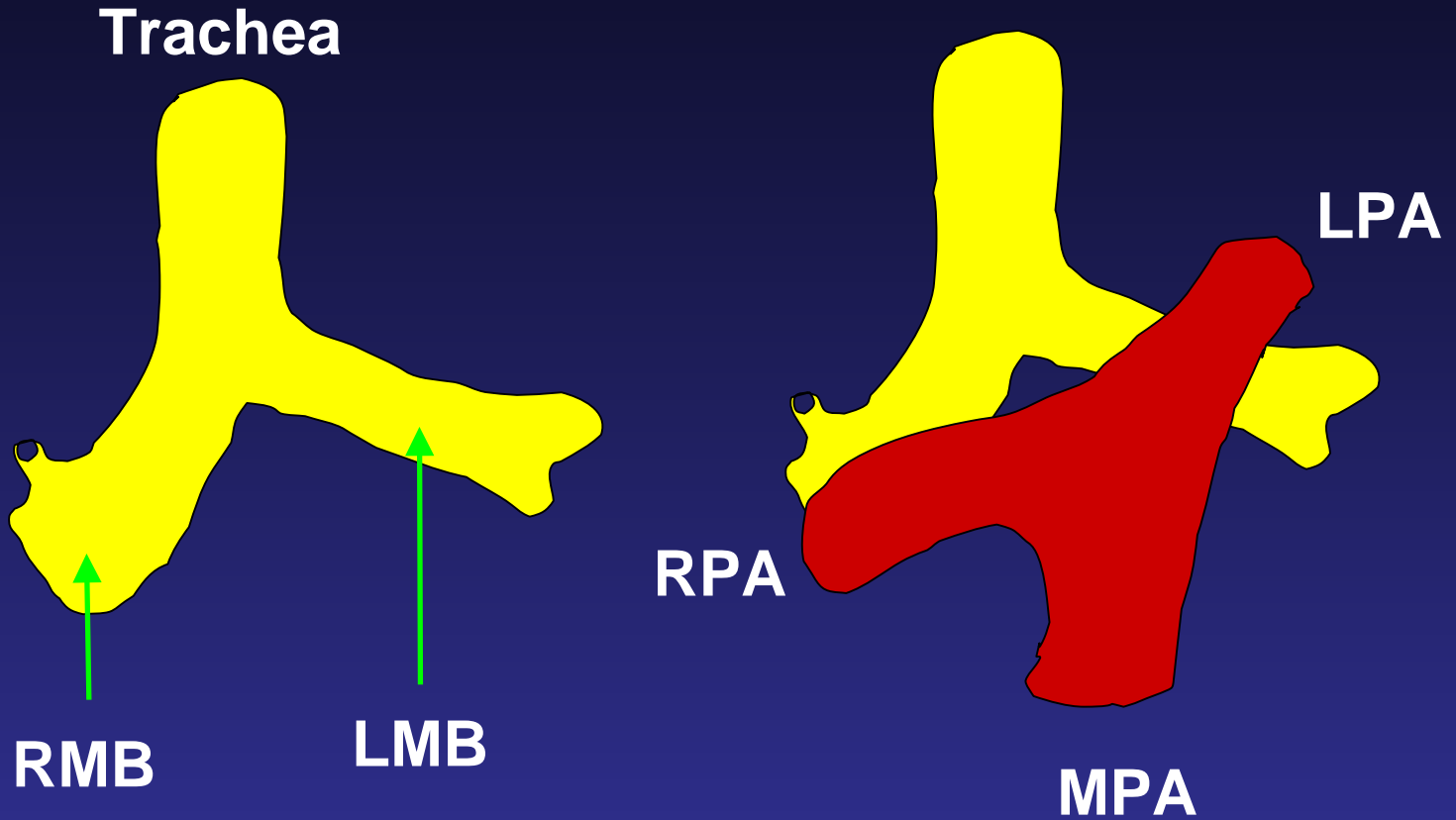
Right atrium



Left atrium



Bronchi and Pulmonary Arteries



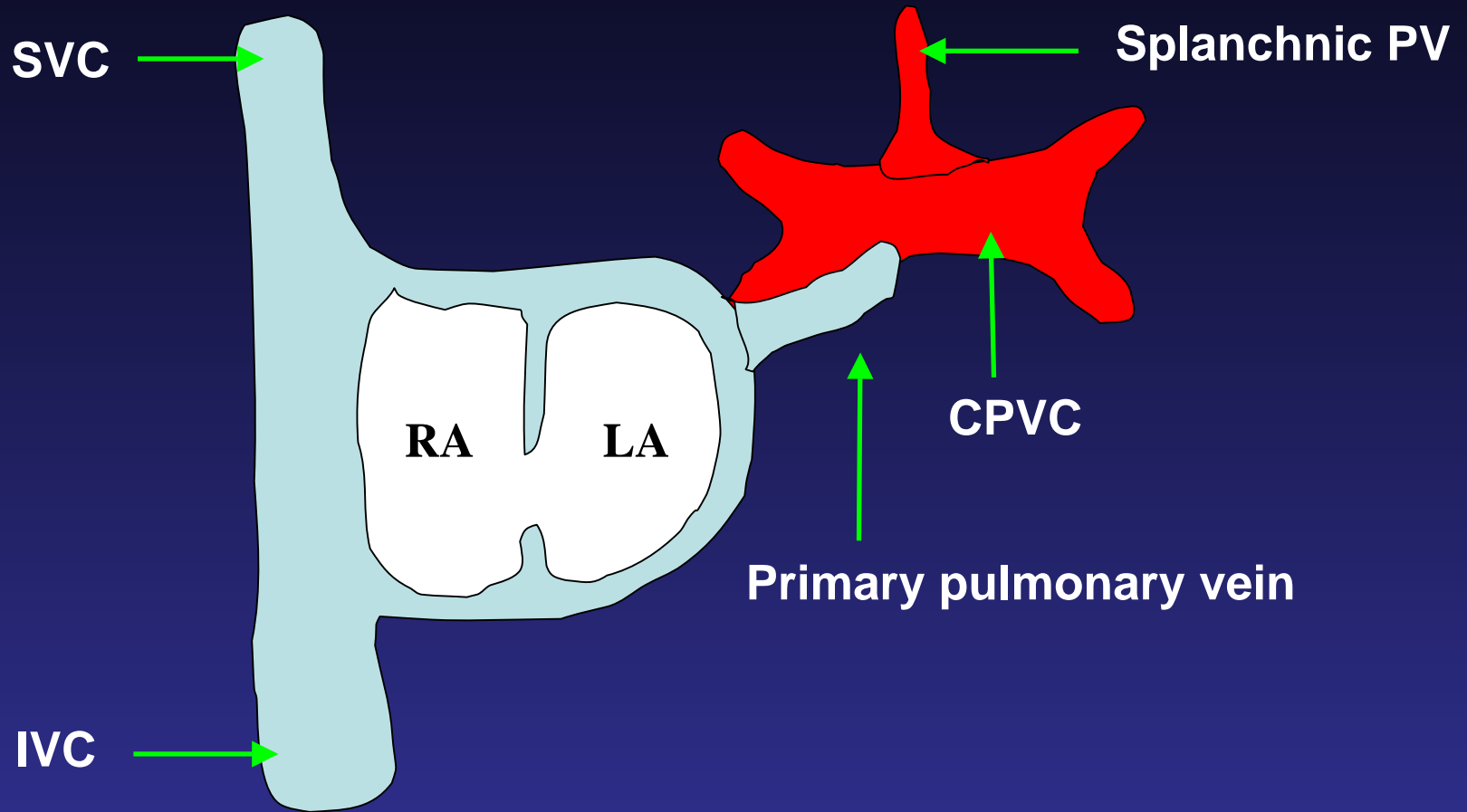
Characteristics of RA and LA

	RA	LA
Auricle Shape	Blunt tip, triangular	Sharp tip, finger-like
Muscle	Abundant	Scanty
Crista terminalis	+	-
Septum	Fossa ovalis	Smooth
SA node	+	-
SVC	?	?
IVC	+	-
PV	-	+
Ipsilateral lung	3 lobes	2 lobes
Ipsilateral bronchus	Eparterial bronchus	Hyparterial bronchus

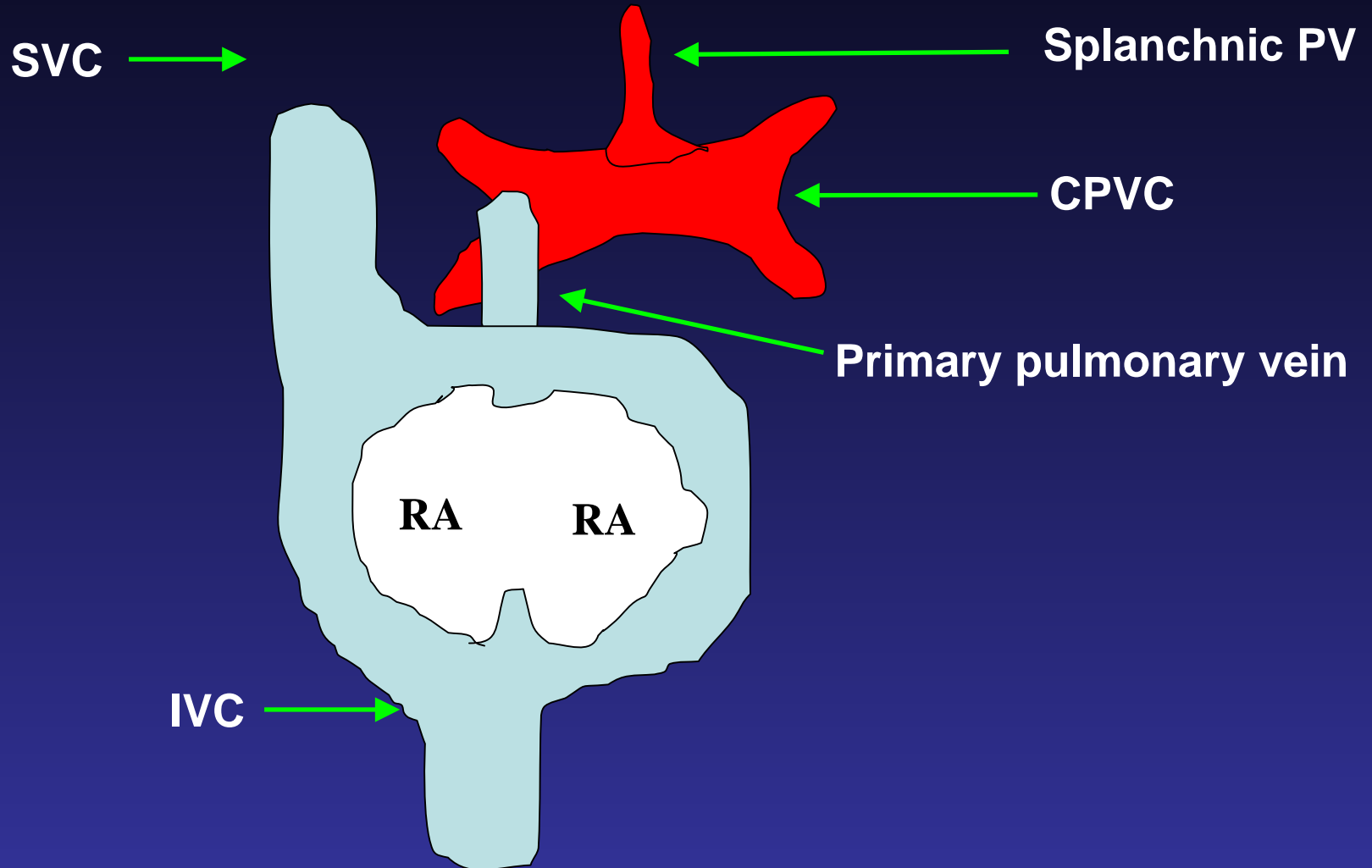
Characteristics of Isomerism

	<i>Rt.isomerism</i>	<i>Lt.isomerism</i>
Auricle	bilateral RA	bilateral LA
Crista terminalis	bilateral	absent
SA node	bilateral	abnormal
SVC	?	?
IVC	Midline (with Ao)	Interruption with azygos continuation
PV	TAPVR	Wide RPV-LPV
Lung	Bilateral 3 lobes	Bilateral 2 lobes
Bronchus	Bilateral eparterial	Bilateral hyparterial
L.M.B / R.M.B	<1.6	<1.6
Spleen	Asplenia	Polysplenia
Px	Poor	Good

Primary pulmonary vein



Primary pulmonary vein in RAI



TAPVD association in RAI

Incidence: 60% - 100%

(Varies according to the definition of TAPVD)

Variables:

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

Objective

Premise:

RAI associated with TAPVD is fatal

Question:

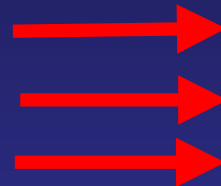
Can we modify the dismal prognosis by aggressive therapeutic strategies, including primary SR?

Patients and Methods

All patients who registered to HSC
between 1994 and 2003

Evolution of Therapeutic Strategies

Treatment withdrawal
No repair of TAPVD
Direct repair of TAPVD
No ECMO / VAD
Medical Tx of arrhythmia



No Treatment withdrawal
SR for TAPVD repair
ECMO / VAD
Surgical Tx of arrhythmia

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

Patient profile

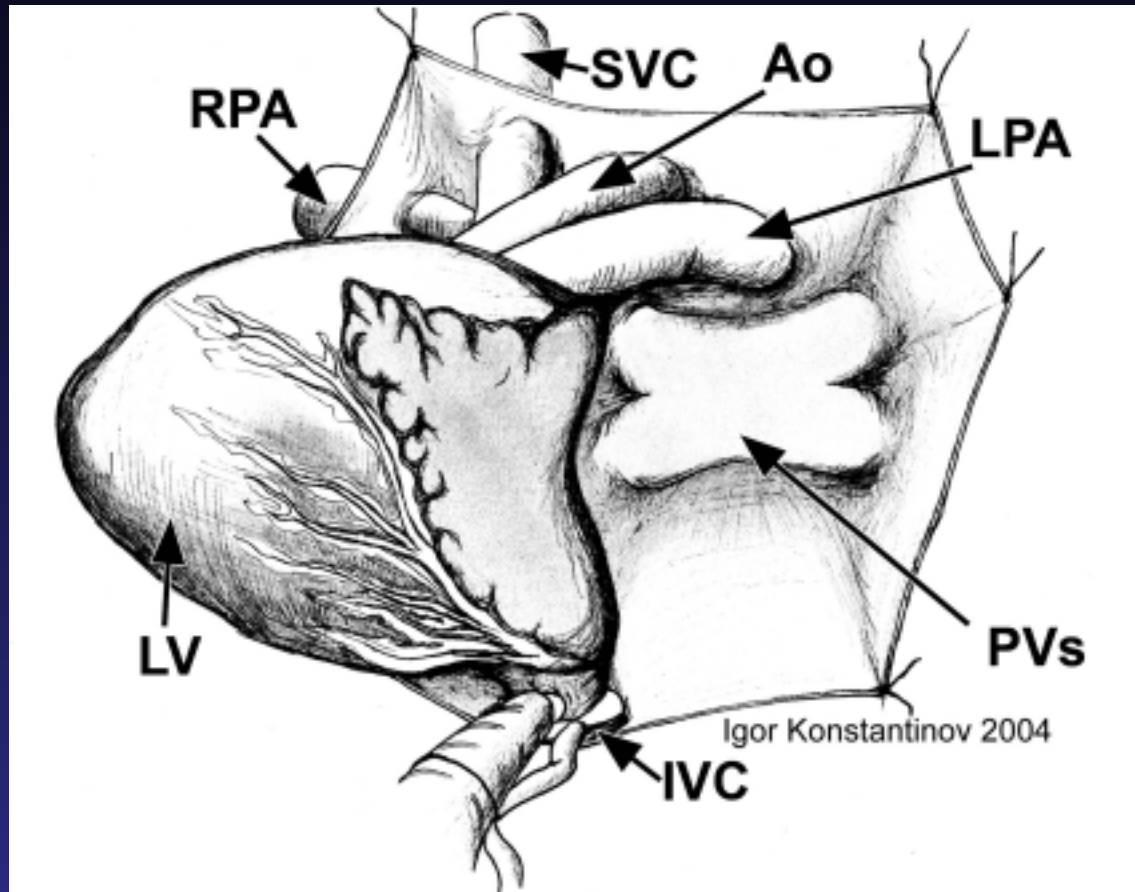
- **Fifty-five Children of RAI**
- **Median age of initial visit: 2 days**
- **TAPVD : 51/55 (93%)**
 - **Extra-cardiac TAPVD: 42/55 (76%)**
 - **Obstructive TAPVD: 22/55 (40%)**
- **Treatment Withdrawal : 11/55 (20%)**
- **Surgical Management: 44/55 (80%)**
 - **Without TAPVD repair: 13/44**
 - **With TAPVD repair: 31/44**

Types of TAPVD (n=55)

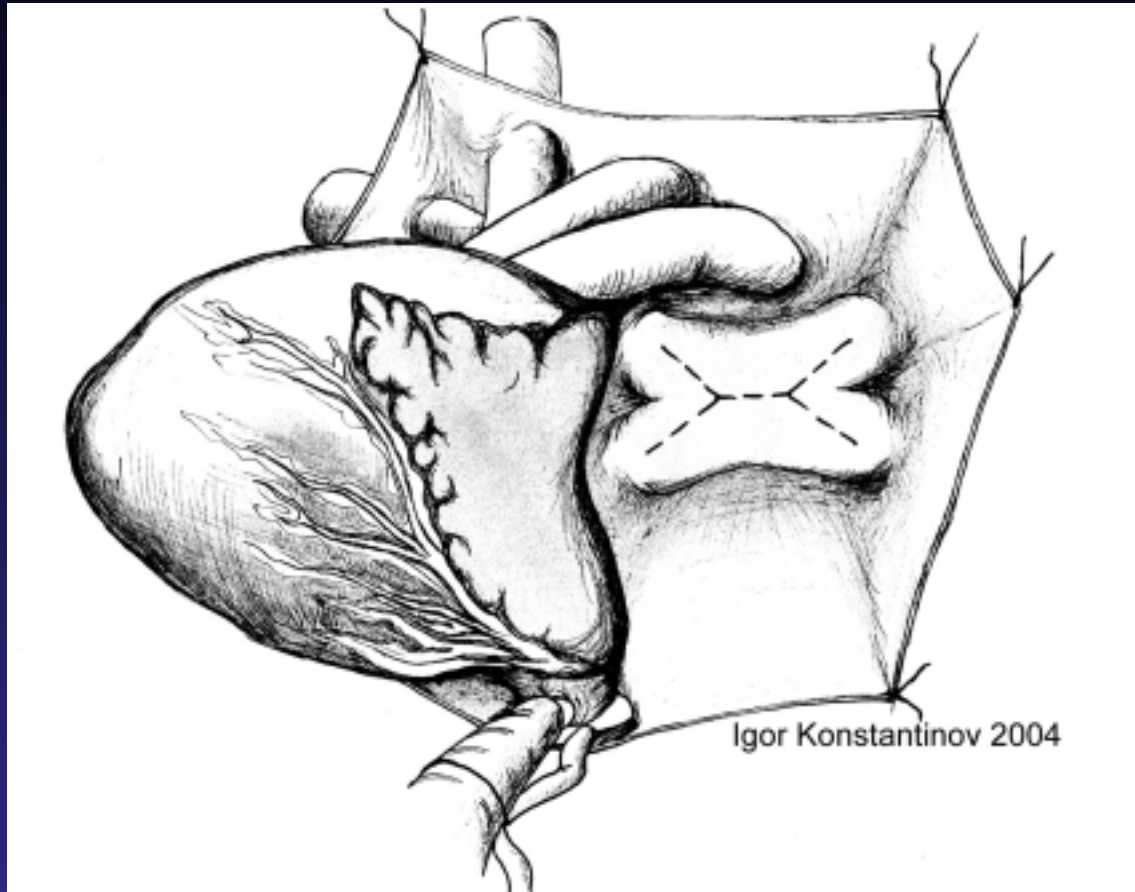
Group	No TAPVD	Supracardiac	Cardiac	Infraracardiac	Mixed
No Treatment	0	5(2)	1(0)	1(1)	4(3)
TAPVD repair (-)	4(0)	3(1)	3(0)	0	3(0)
TAPVD repair (+)	0	19(7)	5(4)	5(3)	2(1)

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

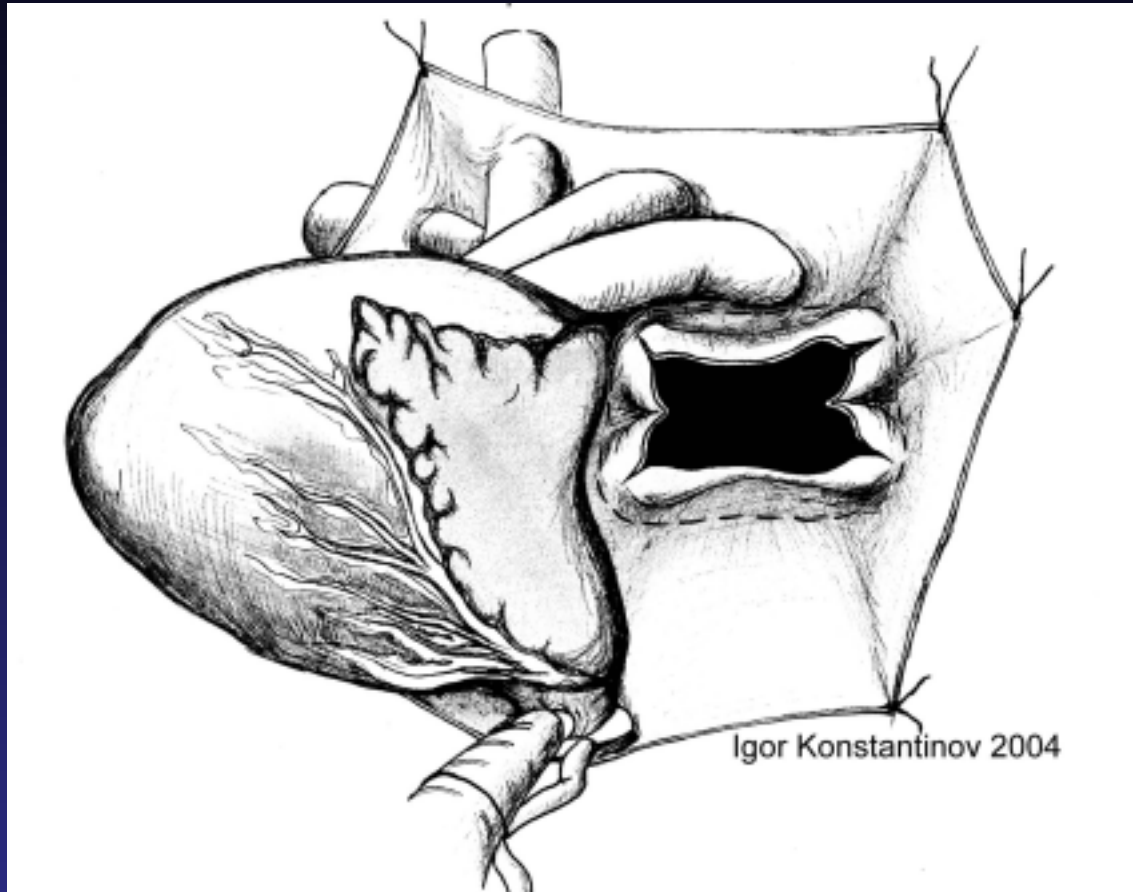
SR without Retro-cardiac adhesion



SR without Retro-cardiac adhesion



SR without Retro-cardiac adhesion



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)

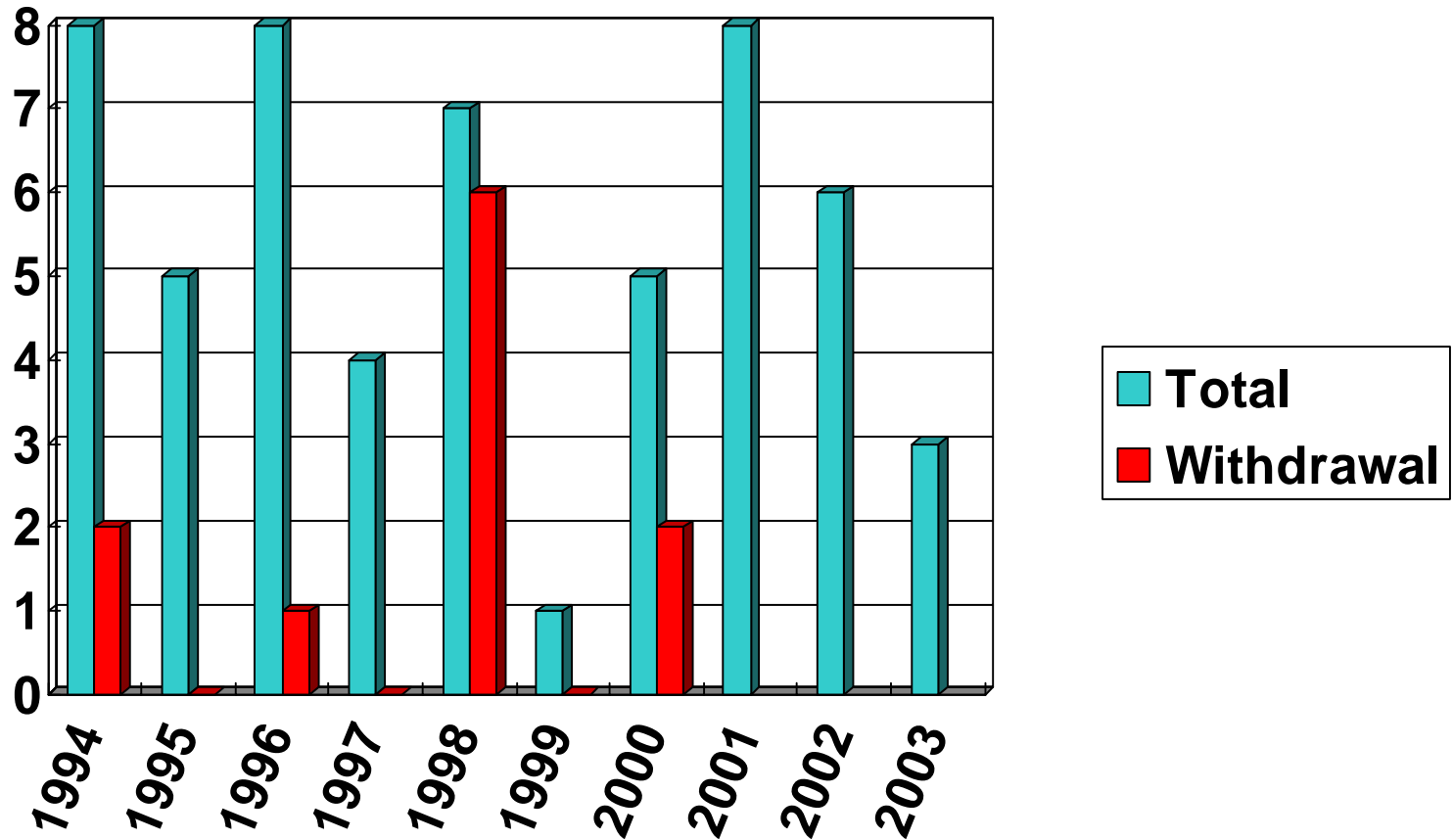
Complex obstructive TAPVD (n=2)

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



Operative procedures

	TAPVD repair (-)	TAPVD repair (+)
Systemic-pulmonary shunt	7	7
PA banding	2	4
BCPS	1	9
Fontan operation	1	3
Cardiac Transplantation	0	2
PA angioplasty	3	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

BVR in general

- **Absolute Contraindication**

1. Unbalanced / Inseparable AV valves
2. Unbalanced / Inseparable Ventricles

- **Relative Contraindication**

1. Complicated systemic and pulmonary venous return
2. Overriding or straddling of atrioventricular valves
3. Complex atrioventricular septal defect
4. Ventriculoarterial connection anomalies

Why is BVR rare in RAI?

- High early mortality of RAI
- AVSD in RAI
- Association of complex VA connection

55 patients

**Withdrawal of Tx
(n=11)**

**No TAPVD repair
(n=13)**

**TAPVD repair
(n=31)**

10

**Death
(n=10)**

1

**Survival without further Tx
(n=1)**

55 patients

**Withdrawal of Tx
(n=11)**

**No TAPVD repair
(n=13)**

**TAPVD repair
(n=31)**

**BCPS
(n=5)**

**Death
(n=10)**

**Fontan
(n=3)**

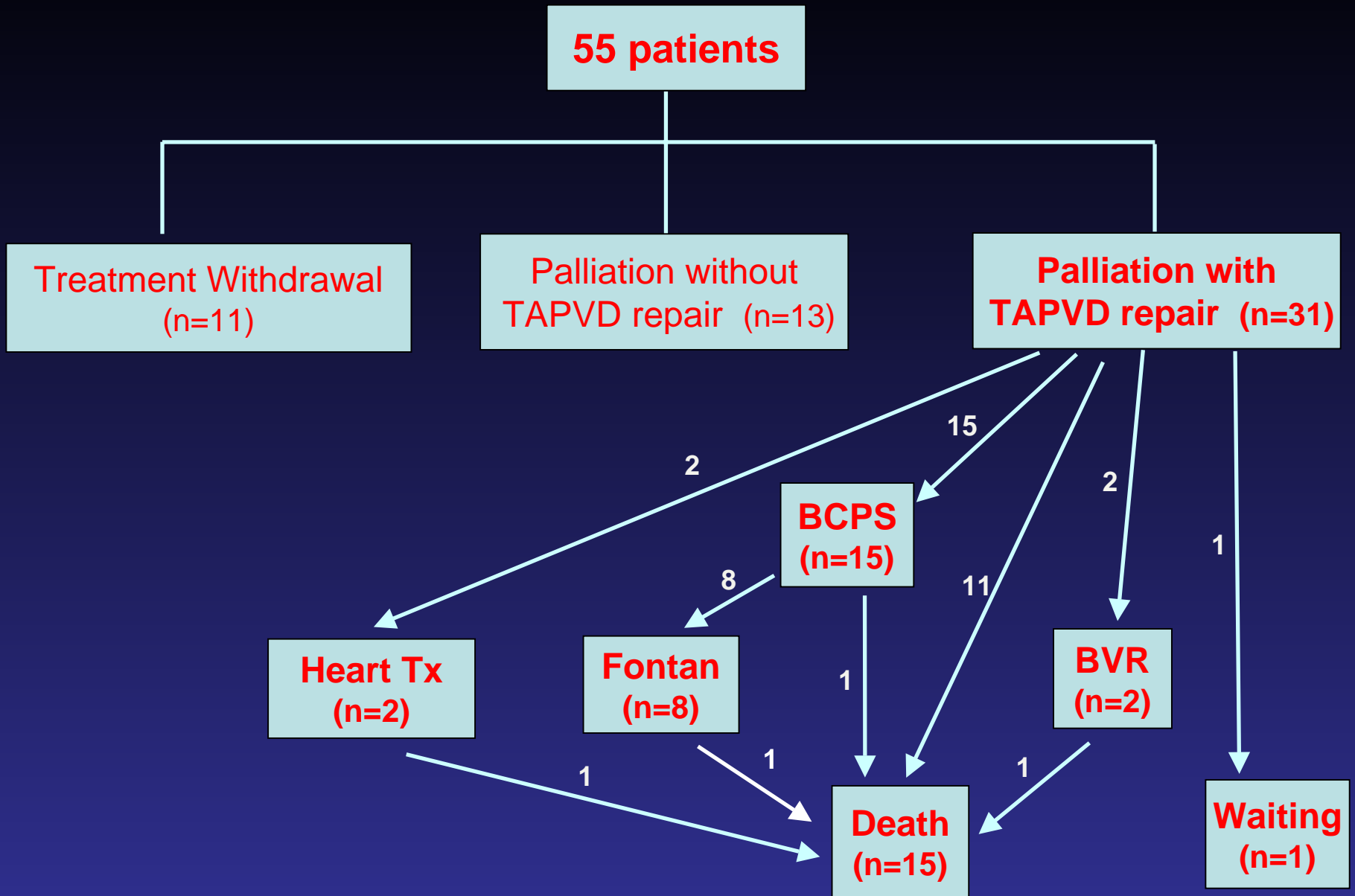
9

3

1

1

2



ECMO / VAD support in RAI

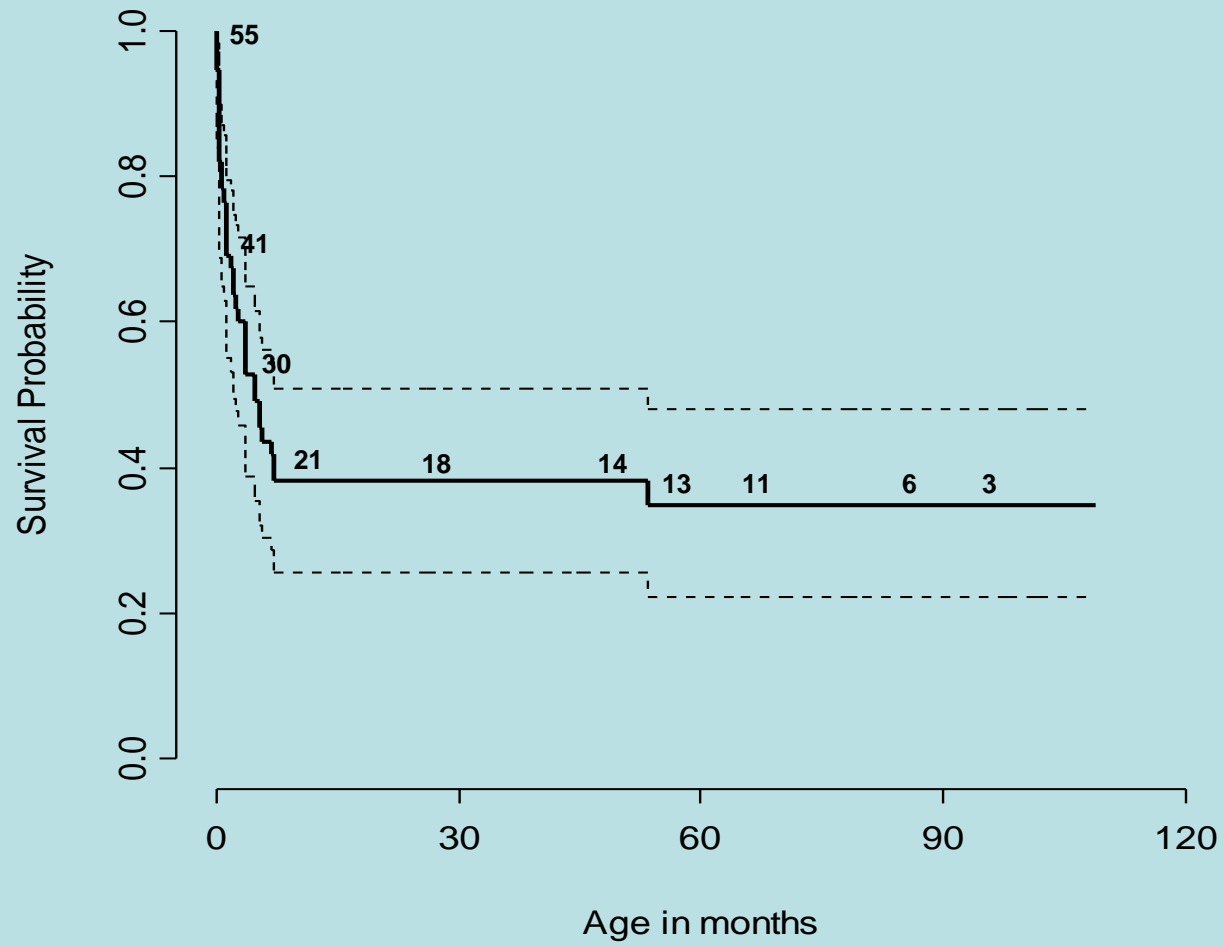
- Rationale: **control of**
 1. post-op pulmonary vascular reactivity
 2. Medically intractable arrhythmia
- ECMO / VAD : 2 / 2
- Successful coming-off: 3 / 4
- Successful discharge: 3 / 3
- Long-term survivor: 1 / 3

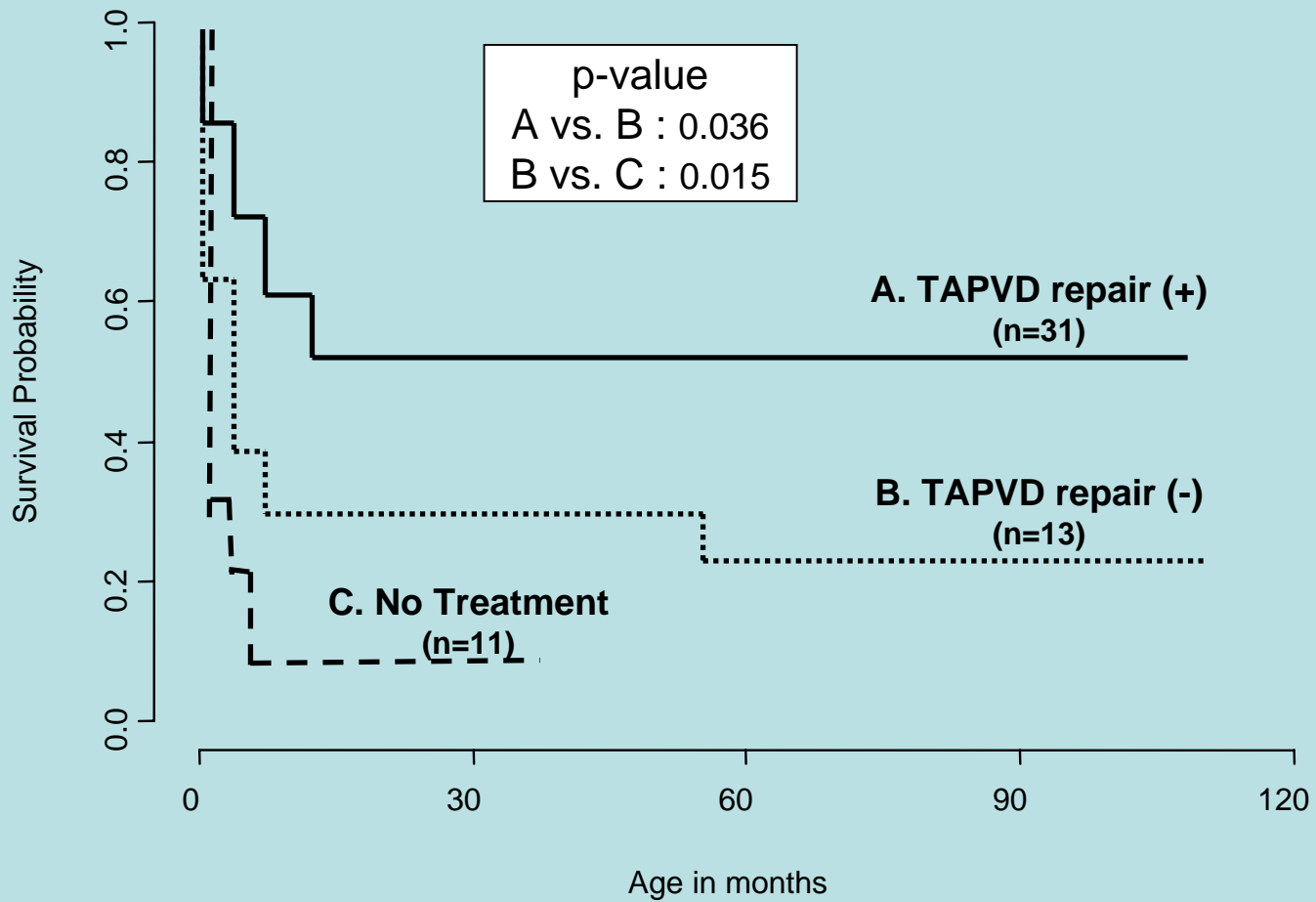
Arrhythmia issue in RAI

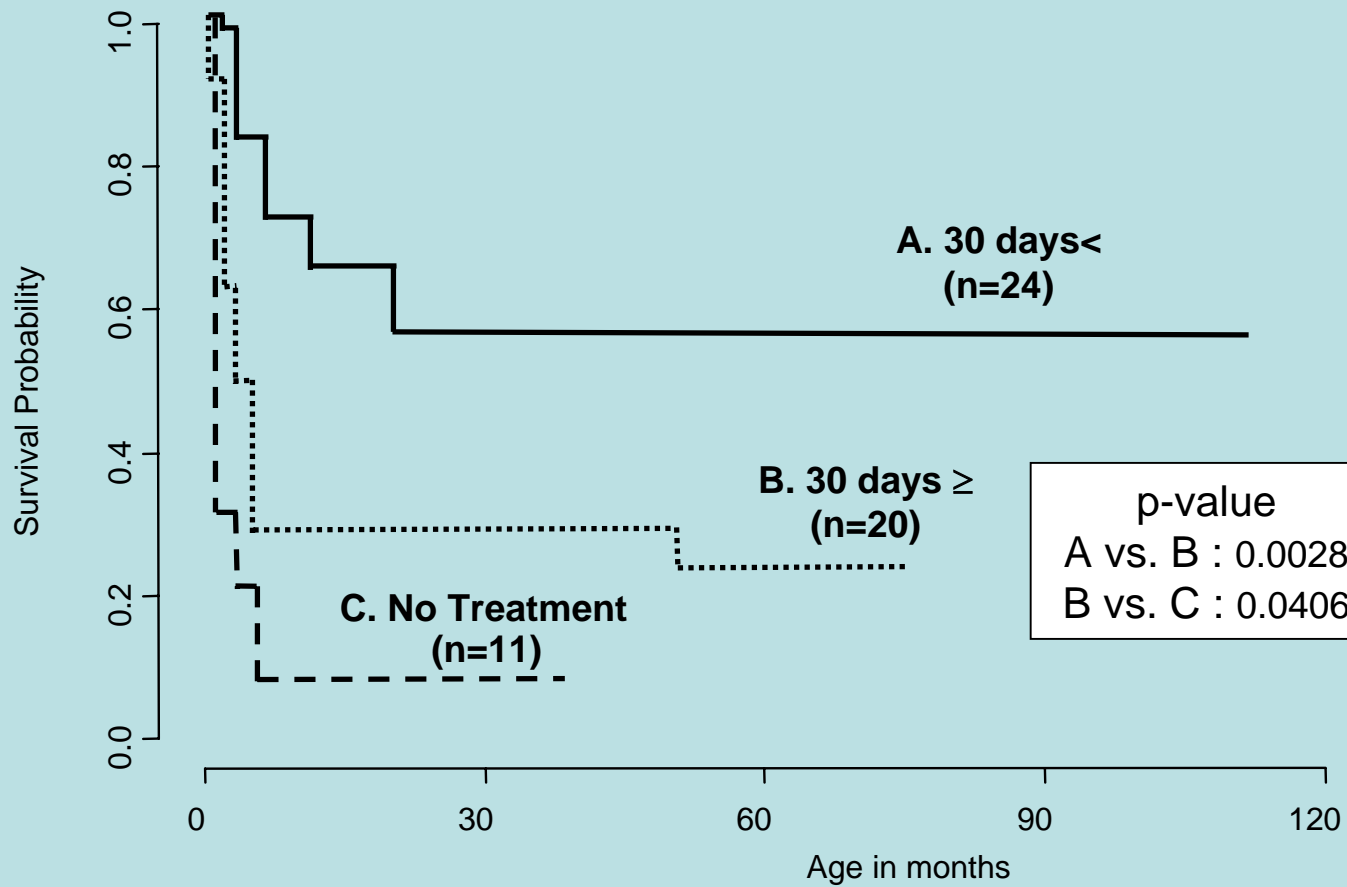
- **Medical treatment**
- **Medically uncontrollable PSVT**
ECMO / VAD
- **Surgical ablation if needed**

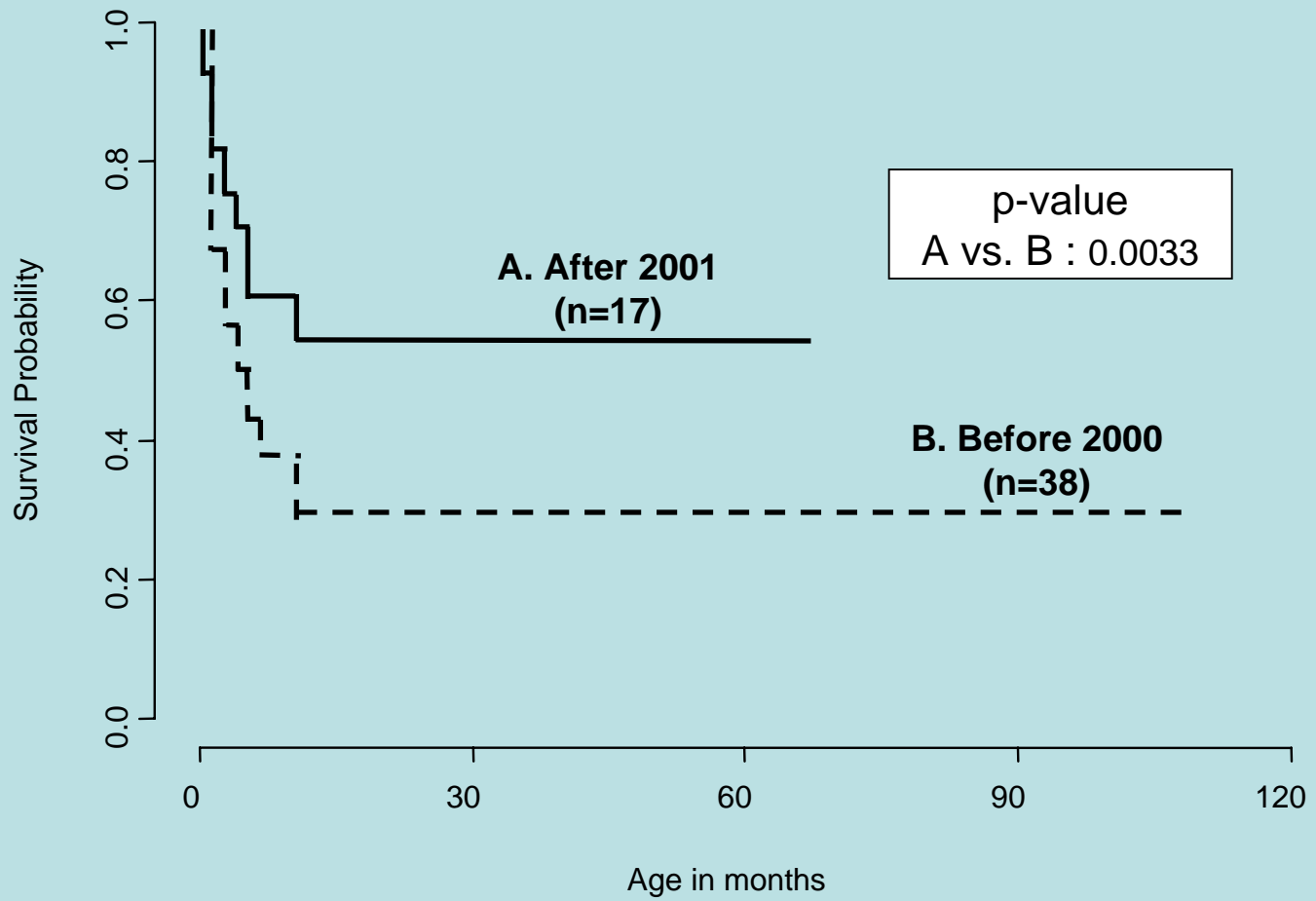
Results

- **Two risk factors of decreased survival**
 - 1) Obstructive TAPVD
 - 2) Mixed and infracardiac type TAPVR
- **After adjustment, primary sutureless repair of TAPVD was associated with improved survival**
(Hazard ratio=0.43, p=0.19)









Conclusions

- **Outcome of RAI continue to be less than Optimal in current series.**
- **Contemporary therapeutic strategy appeared to have a positive impact on survival**
- **Role of primary SR for TAPVD need further evaluation.**