Chirurgie de l'Hypertension Pulmonaire

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Université Paris Sud

- Endartériectomie pulmonaire
- Transplantation
- Assistance cardio-respiratoire
- Traitements palliatifs
- Vers un poumon artificiel?

1 Pulmonary arterial hypertension (PAH)

- 1.1 Idiopathic
- 1.2 Heritable
  - 1.2.1 BMPR2
  - 1.2.2 ALK1, endoglin (with or without hereditary haemorrhagic telangiectasia)
  - 1.2.3 Unknown
- 1.3 Drugs and toxins induced
- 1.4 Associated with (APAH)
  - 1.4.1 Connective tissue diseases
  - 1.4.2 HIV infection
  - 1.4.3 Portal hypertension
  - 1.4.4 Congenital heart disease
  - 1.4.5 Schistosomiasis
  - 1.4.6 Chronic haemolytic anaemia
- 1.5 Persistent pulmonary hypertension of the newborn

1′ Pulmonary veno-occlusive disease and/or pulmonary capillary haemangiomatosis

2 Pulmonary hypertension due to left heart disease

- 2.1 Systolic dysfunction
- 2.2 Diastolic dysfunction
- 2.3 Valvular disease

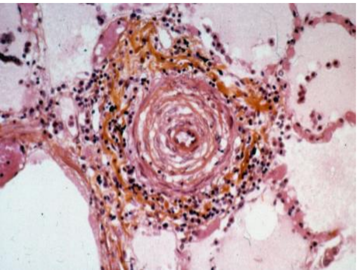
3 Pulmonary hypertension due to lung diseases and/or hypoxia

- 3.1 Chronic obstructive pulmonary disease
- 3.2 Interstitial lung disease
- 3.3 Other pulmonary diseases with mixed restrictive and obstructive pattern
- 3.4 Sleep-disordered breathing
- 3.5 Alveolar hypoventilation disorders
- 3.6 Chronic exposure to high altitude
- 3.7 Developmental abnormalities

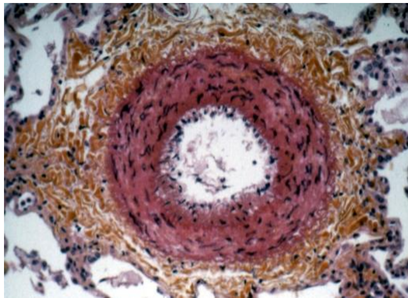
4 Chronic thromboembolic pulmonary hypertension

5 PH with unclear and/or multifactorial mechanisms

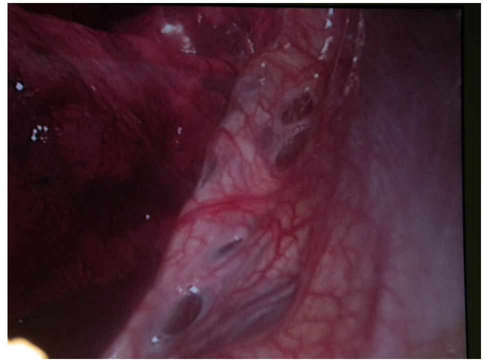
- 5.1 Haematological disorders: myeloproliferative disorders, splenectomy.
- 5.2 Systemic disorders: sarcoidosis, pulmonary Langerhans cell histiocytosis, lymphangioleiomyomatosis, neurofibromatosis, vasculitis
- 5.3 Metabolic disorders: glycogen storage disease, Gaucher disease, thyroid disorders
- 5.4 Others: tumoural obstruction, fibrosing mediastinitis, chronic renal failure on dialysis

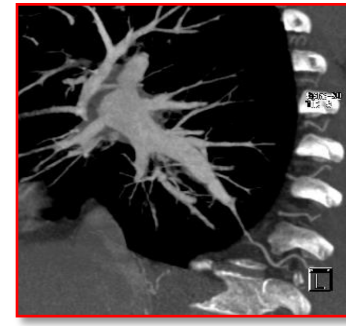


Classification Clinique de l'Hypertension Pulmonaire

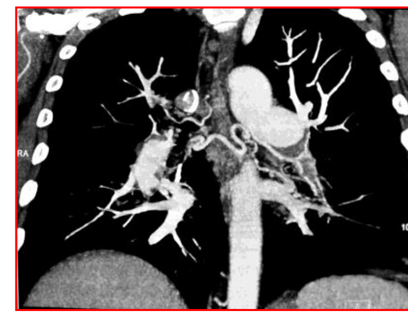


G Simonneau et al JACC 2009

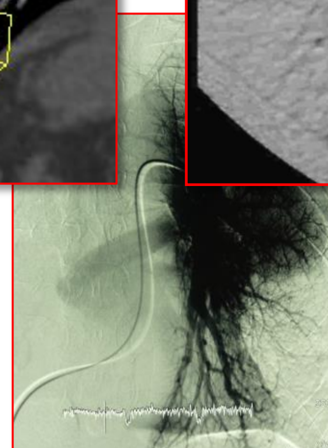


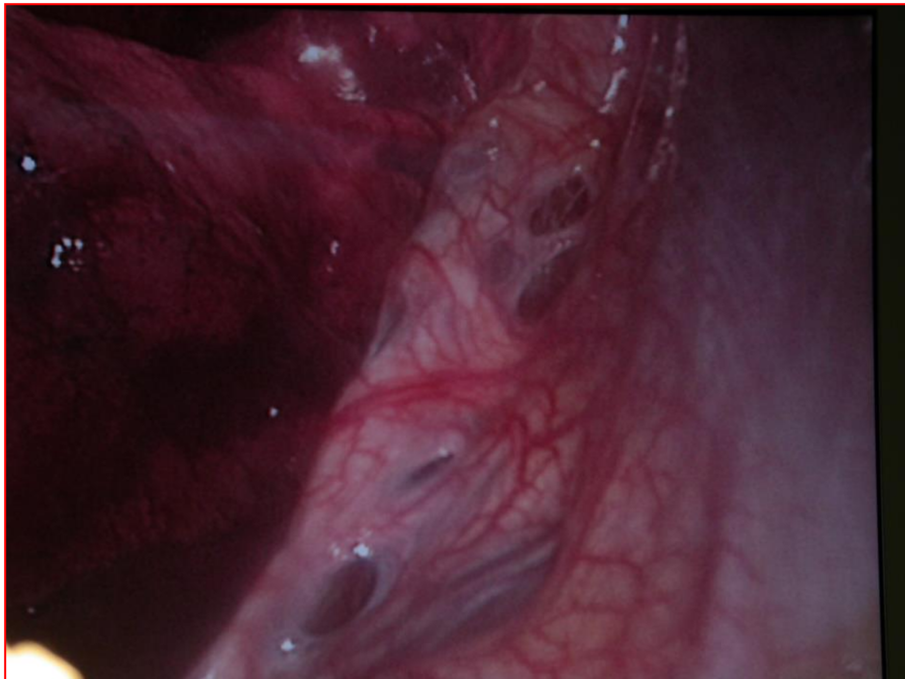


HP-PE

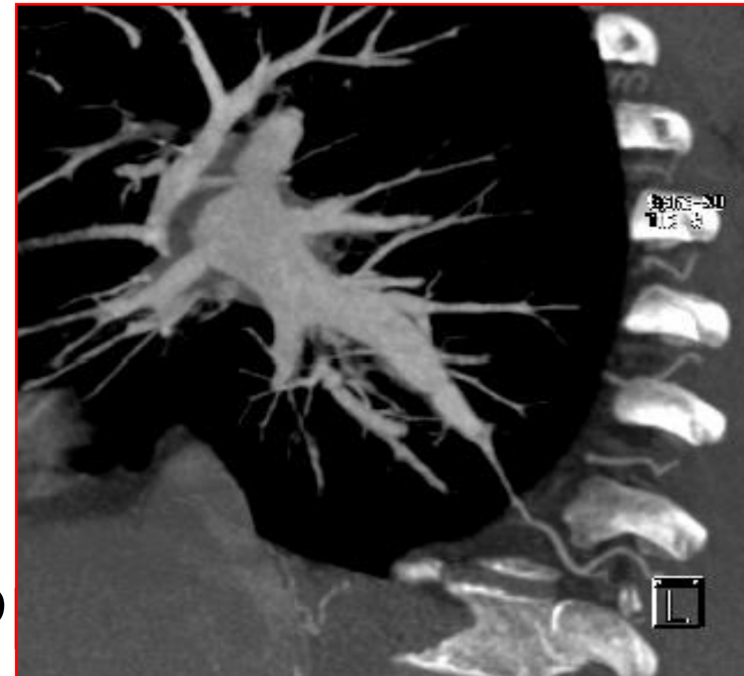


HP

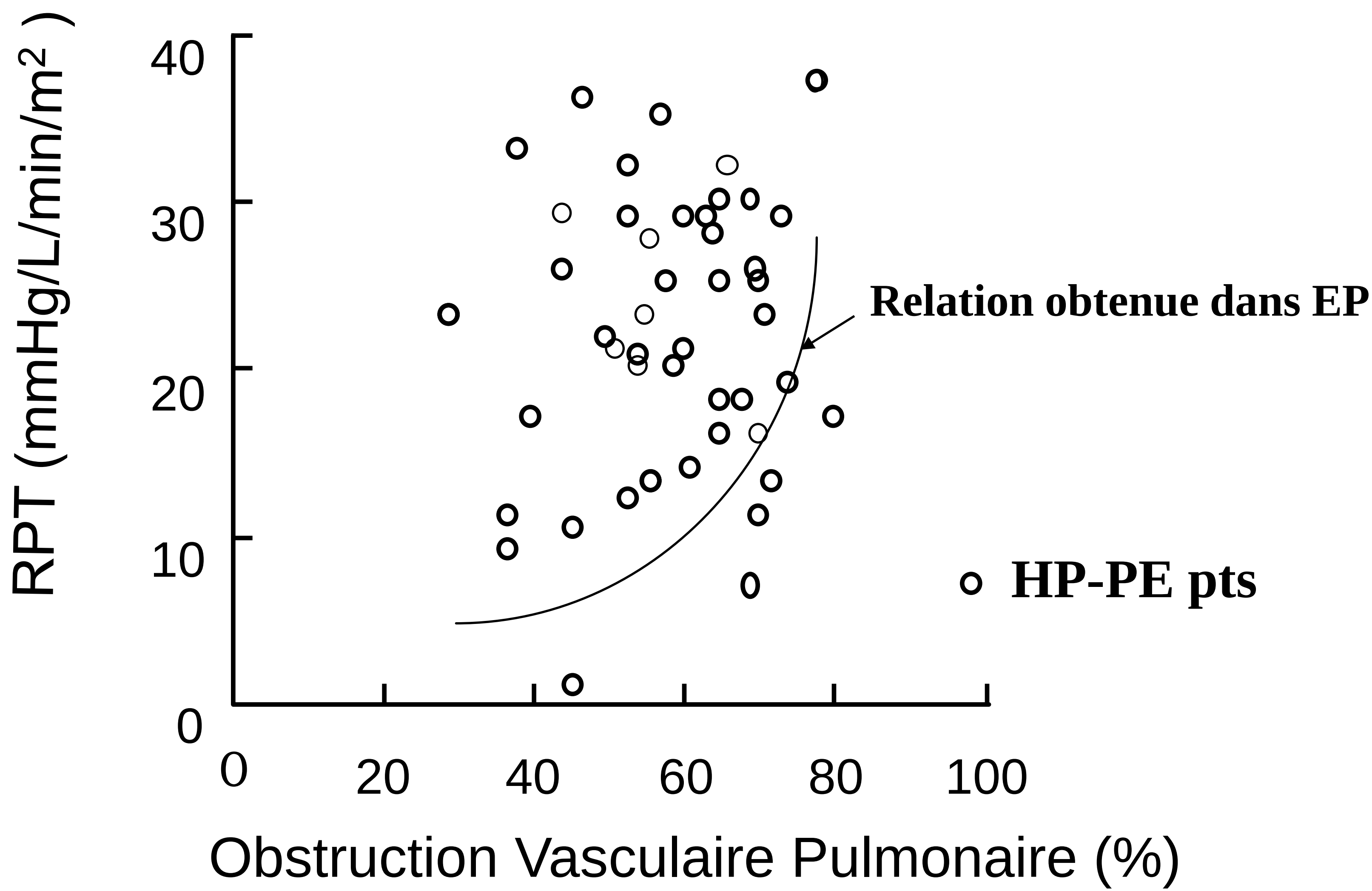




P-P

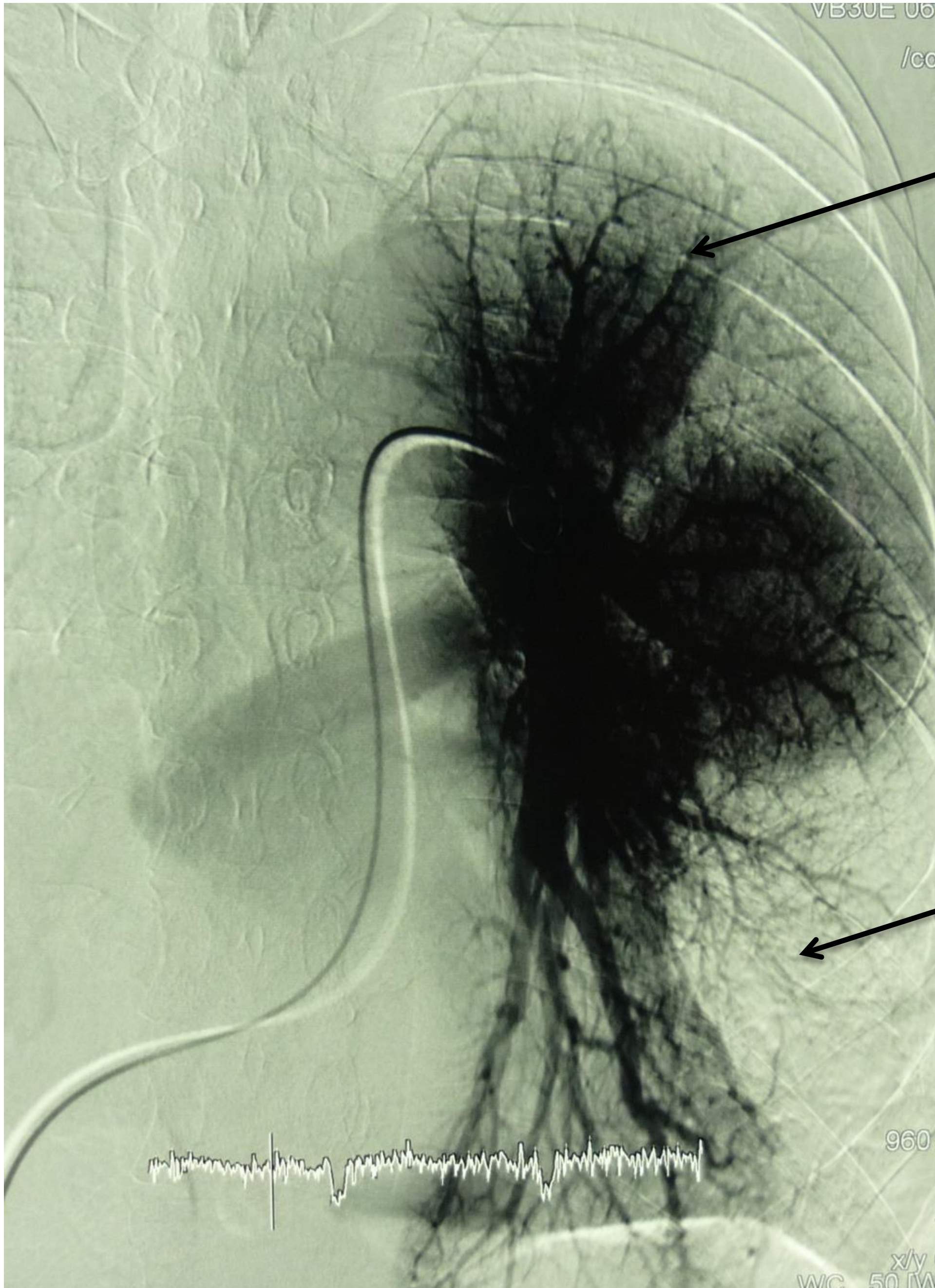


Atteinte microvasculaire



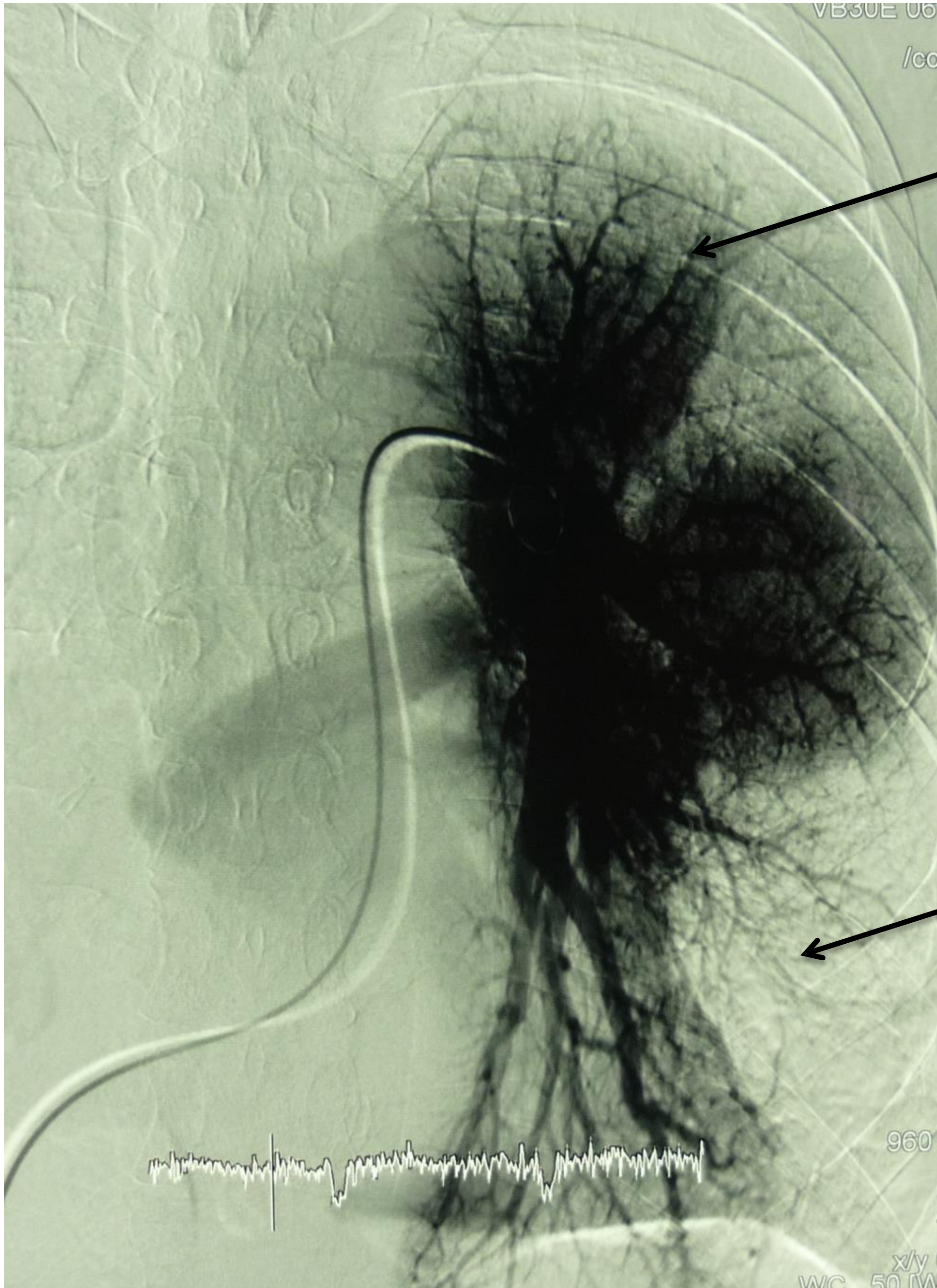
Azarian et al, J Nucl Med, 1997

TERRITOIRES NON OBSTRUES



TERRITOIRES NON OBSTRUES

TERRITOIRES OBSTRUES



TERRITOIRES NON OBSTRUES

VASCULOPATHIE PULMONAIRE DISTALE

TERRITOIRES OBSTRUES

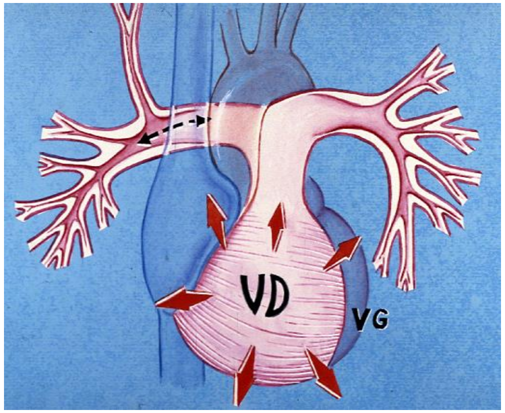
ISCHEMIE CHRONIQUE VASCULOPATHIE POST OBSTRUCTIVE

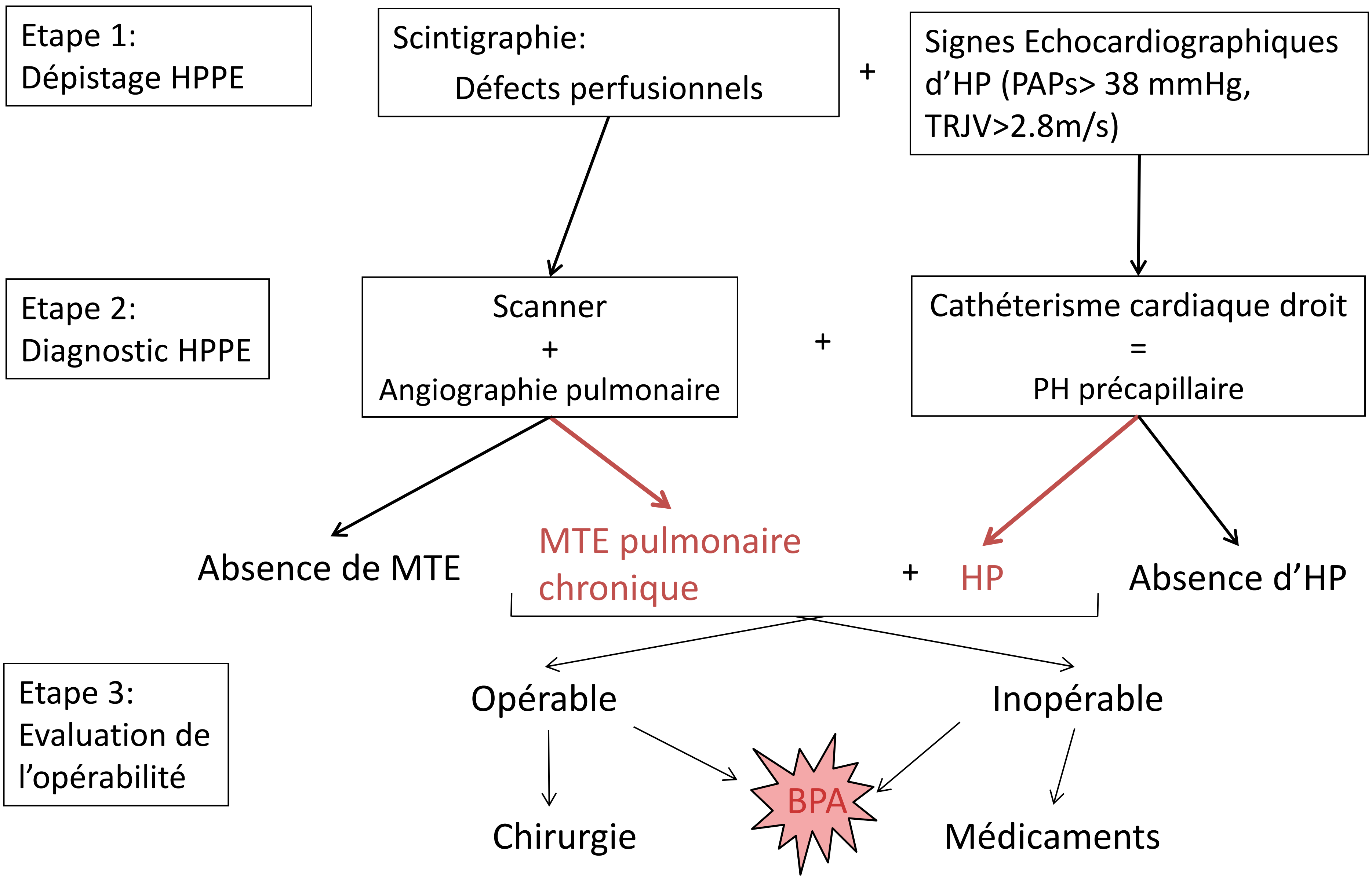
Présentation Clinique

- Nouvel épisode d'une embolie pulmonaire aiguë
- Période de stabilité dite "lune de miel"
- Dyspnée d'aggravation progressive

  Parfois associée à une hémoptysie, douleur thoracique, fatiguabilité, embolies paradoxales

- En urgence devant une défaillance cardiaque droite




Etape 1:
Dépistage HPPE
Scintigraphie:
Défects perfusionnels
+
Signes Echocardiographiques d'HP (PAPs> 38 mmHg, TRJV>2.8m/s)
Etape 2:
Diagnostic HPPE
Scanner
+
Angiographie pulmonaire
+
Cathéterisme cardiaque droit
=
PH précapillaire
Absence de MTE
MTE pulmonaire chronique
+
HP
Absence d'HP
Etape 3:
Evaluation de l'opérabilité
Opérable
Inopérable
Chirurgie
BPA
Médicaments

Indications chirurgicales

- Diagnostic ferme d'HPPE
- Absence de comorbidités excessives
- Matériel endovasculaire accessible (>lobaire)

Facteurs prédictifs de succès

- Antécédent d'EP et de phlébite
- Période lune de miel
- Maladie proximale à l'angiographie
- Circulation systémique hypertrophiée
- Obstruction anatomique corrélée aux résistances

Facteurs prédictifs d’un échec

- Absence d’antécédents d’EP ou de phlébite
- Présence d’un cathéter
- Maladie distale à l’angiographie
- Discordance entre obstruction et sévérité hémodynamique
- Pas de développement de la circulation bronchique
- Splénectomie (Maladie hématologique+++), Maladies inflammatoires chroniques

Endartériectomie pulmonaire

Buts :

- Amélioration hémodynamique en réduisant la post-charge du ventricule droit
- Amélioration des rapports ventilation/perfusion
- Prévention du développement de la vasculopathie pulmonaire distale

Principes de l'endartériectomie

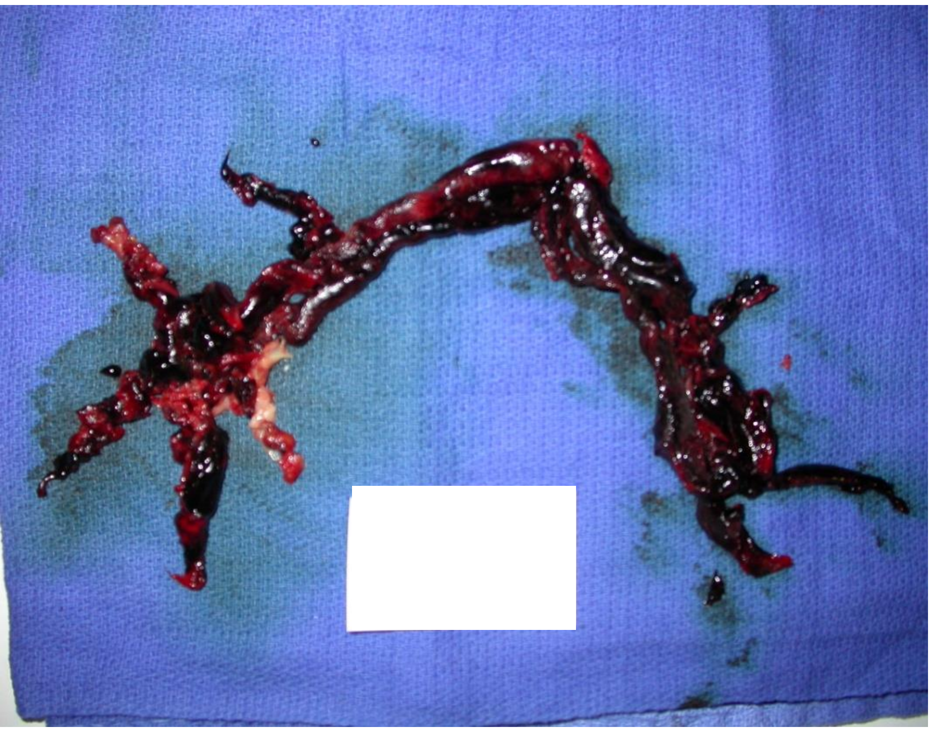
- Procédure bilatérale

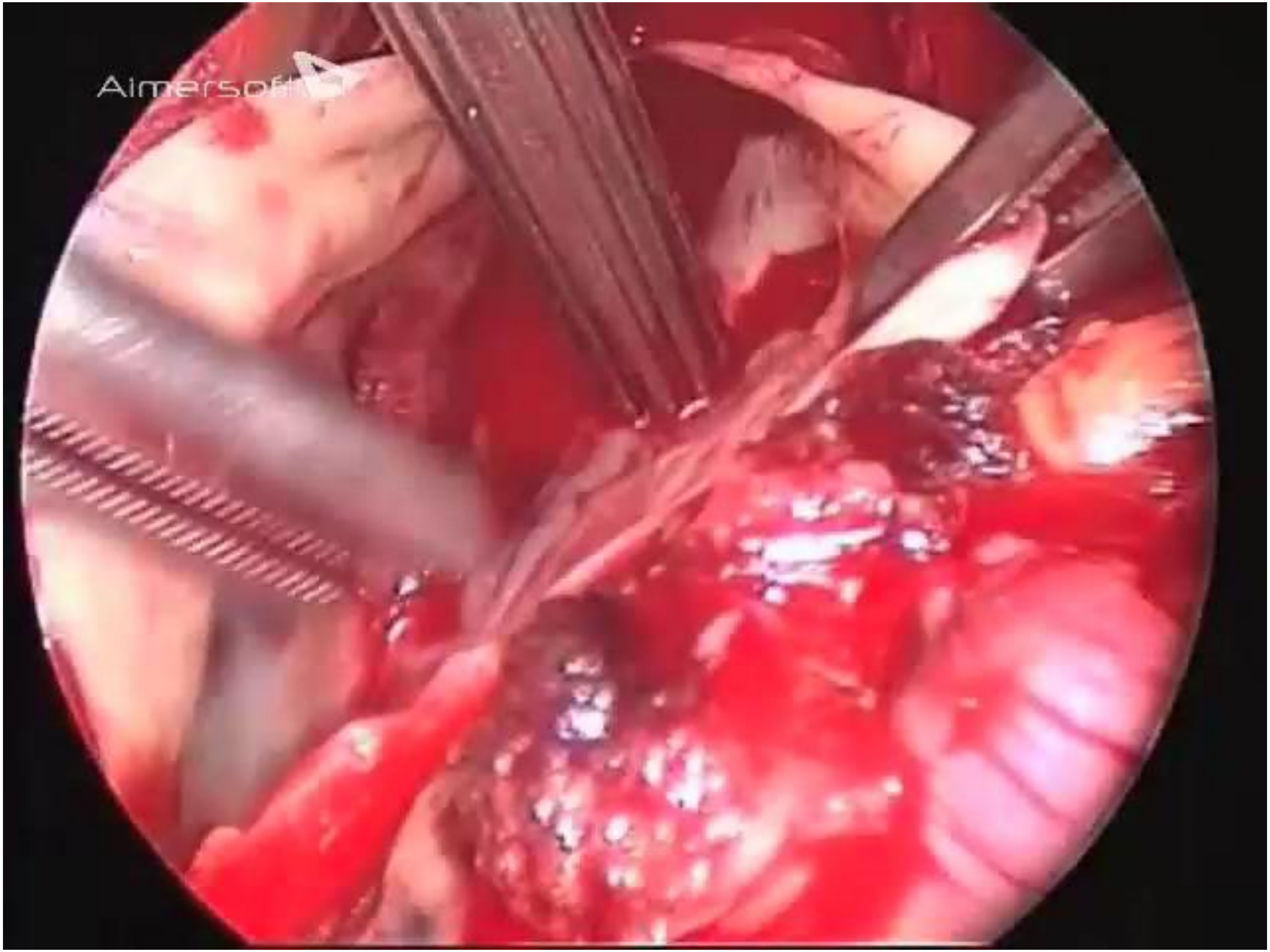
  Enlever au maximum l'obstruction de l'arbre pulmonaire

- Chirurgie intrapéricardique
  - Évite les adhérences pleurales
  - Accès bilatéral
  - Permet l'utilisation de la CEC

- Arrêt circulatoire en hypothermie profonde
  - Arrête le saignement d'origine bronchique et permet une bonne visualisation de l'arbre artériel distal

Endartériectomie pulmonaire




Aimersoft

Expérience de Marie Lannelongue

Janvier 1995 to decembre 2012
n = 1350

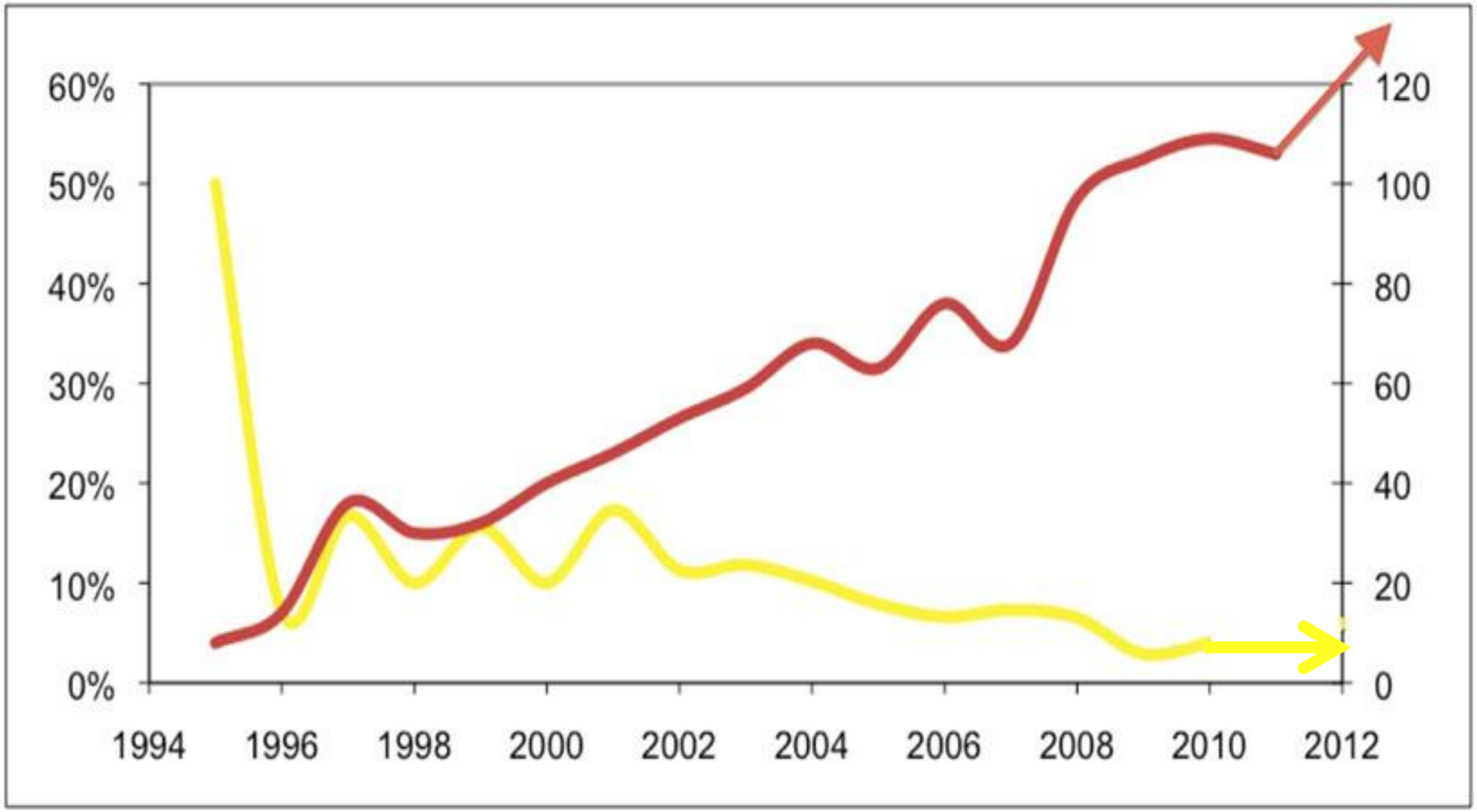
Age (années) :	55 (range 15-83)
PAPm (mm Hg):	54 ± 12.3
Index Cardiaque (L/min/m):	2.2 ± 0.6
RPT (dyne/sec/cm-5):	1206 ± 465

Endartérectomie pulmonaire

n = 1350

Mortalité postopératoire

n/an



2012
N= 139

Mortalité actuelle
2.5%

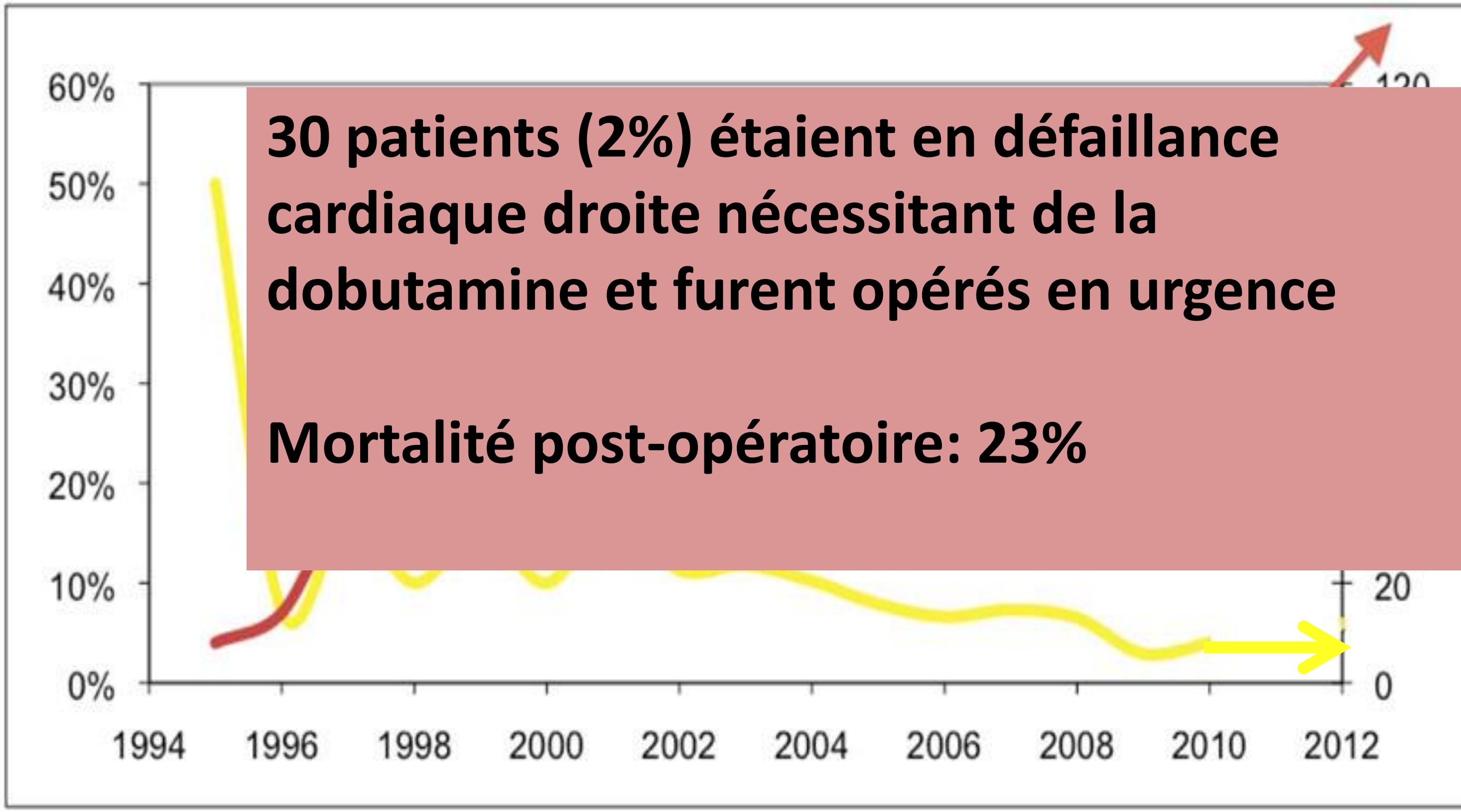
Janvier 1995 to decembre 2012

Endartérectomie pulmonaire

n = 1350

Mortalité
postopératoire

n/an



30 patients (2%) étaient en défaillance cardiaque droite nécessitant de la dobutamine et furent opérés en urgence

Mortalité post-opératoire: 23%

2012
N= 139

Mortalité actuelle
2.5%

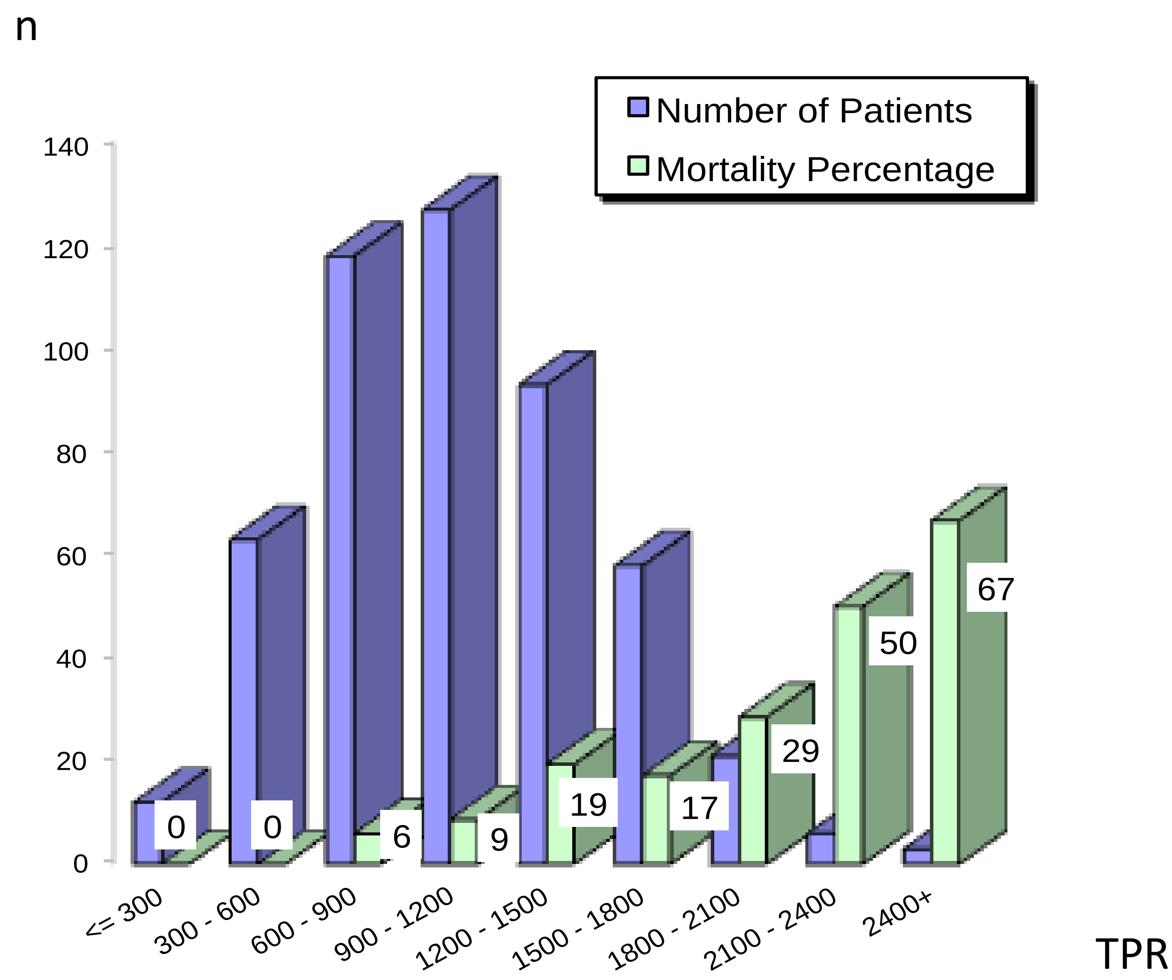
Janvier 1995 to decembre 2012

Mortalité post-opératoire

Mortality increases with preoperative resistance,

p<0,001

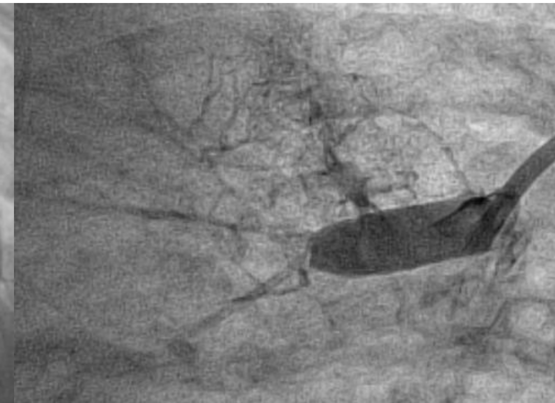
(OR : 1,761 95% CI 1,45 – 2,13)



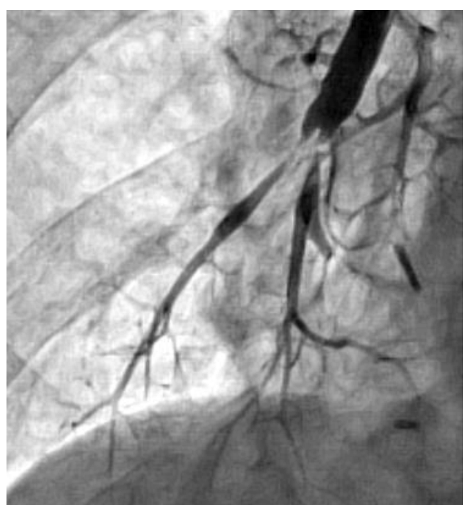
Complications post-opératoires

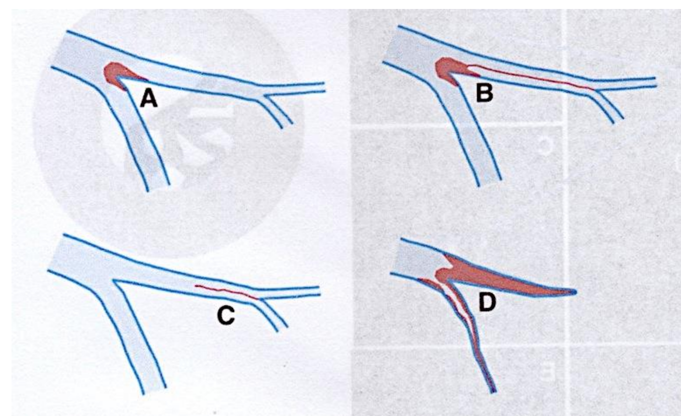
- Oedeme pulmonaire de reperfusion (40 ECLS)
- Défaillance cardiaque droite par hypertension pulmonaire persistante
- Pneumopathie nosocomiale
- Hémoptysies (embolisation n=12)
- Rethrombose (très rare)

STENOSIS



OCCLUSION

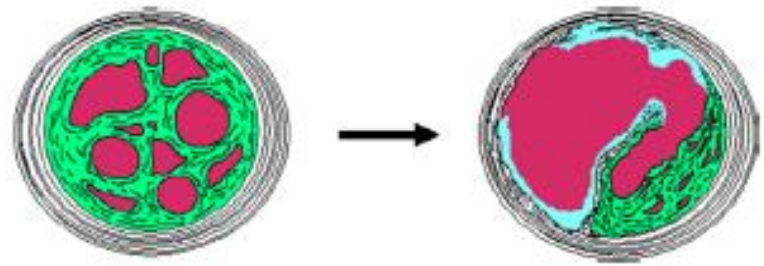


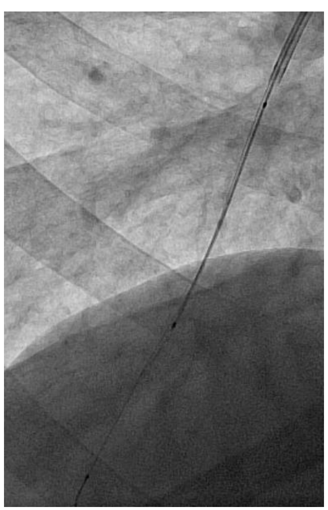


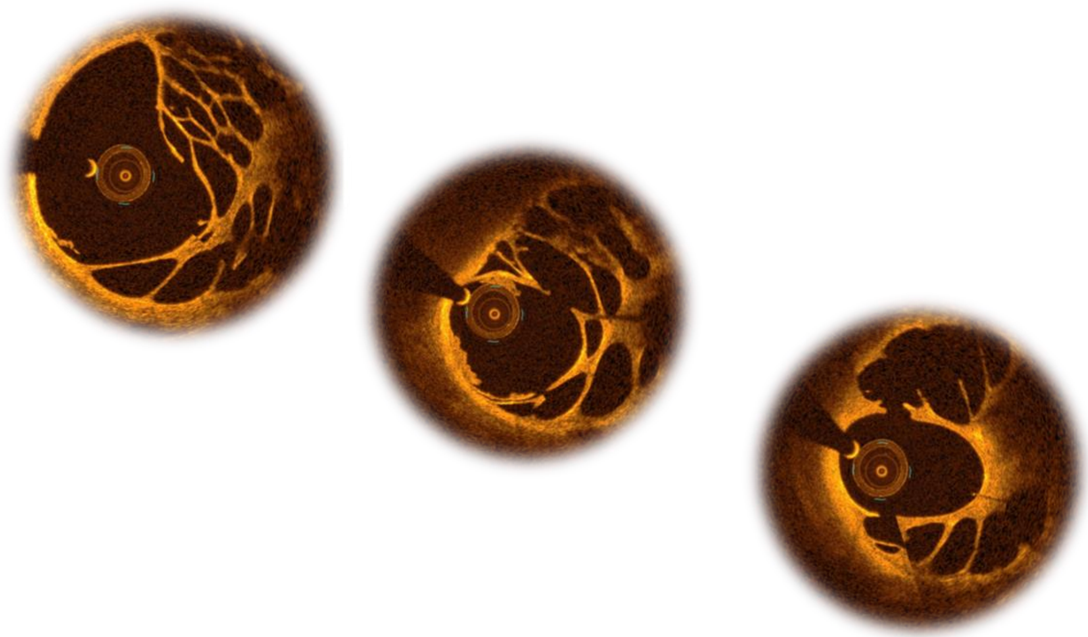
Sugiyama et al
Jpn J Radiol (2014) 75-82

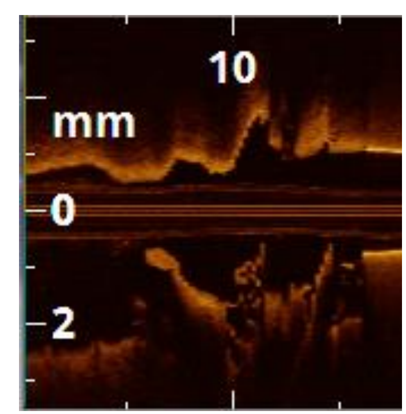
Mécanisme de l'angioplastie pulmonaire

- Dissection dans le plan de la media
- Matériel endovasculaire comprimé sur le côté







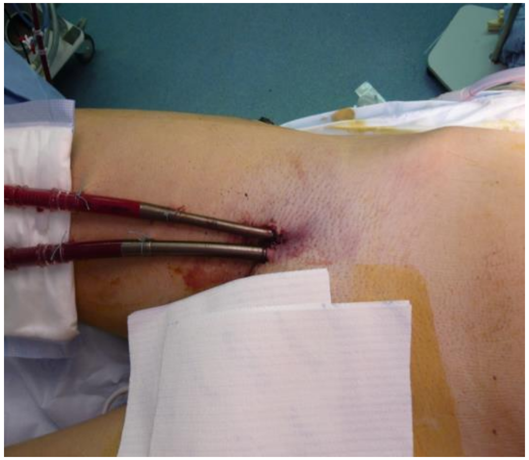


Transplantation : “The Backup plan”

N =11

		Echec précoce	Echec tardif
		8	3
Pont à la Tx		8	0
	AV-ECMO	5	
	Novalung	3	
Type de Tx	HLT	7	0
	DLT	1	3

Mortalité 27% (n=3)
(Rejet aigu 1, DPG 2)



Contents lists available at SciVerse ScienceDirect

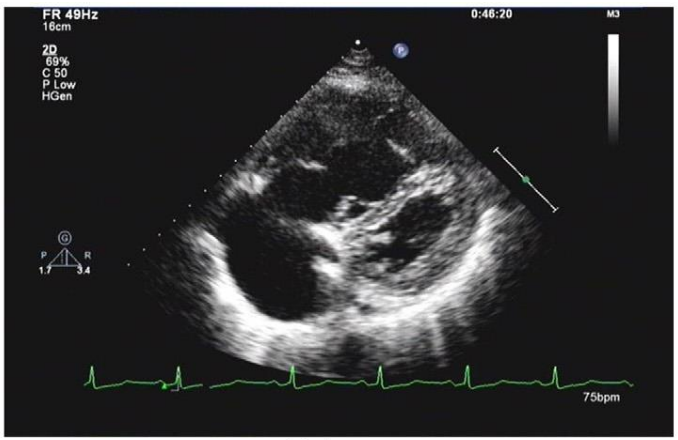
Thrombosis Research

journal homepage: www.elsevier.com/locate/thromres

Regular Article

Improvement of right ventricular dysfunction after pulmonary endarterectomy in patients with chronic thromboembolic pulmonary hypertension: Utility of echocardiography to demonstrate restoration of the right ventricle during 2-year follow-up

Yi-dan Li [a,1], Zhen-guo Zhai [b,c,1], Ya-feng Wu [a,1], Yuan-hua Yang [b,c], Song Gu [d], Yan Liu [d], Pi-xiong Su [d], Chen Wang [c,e,*]

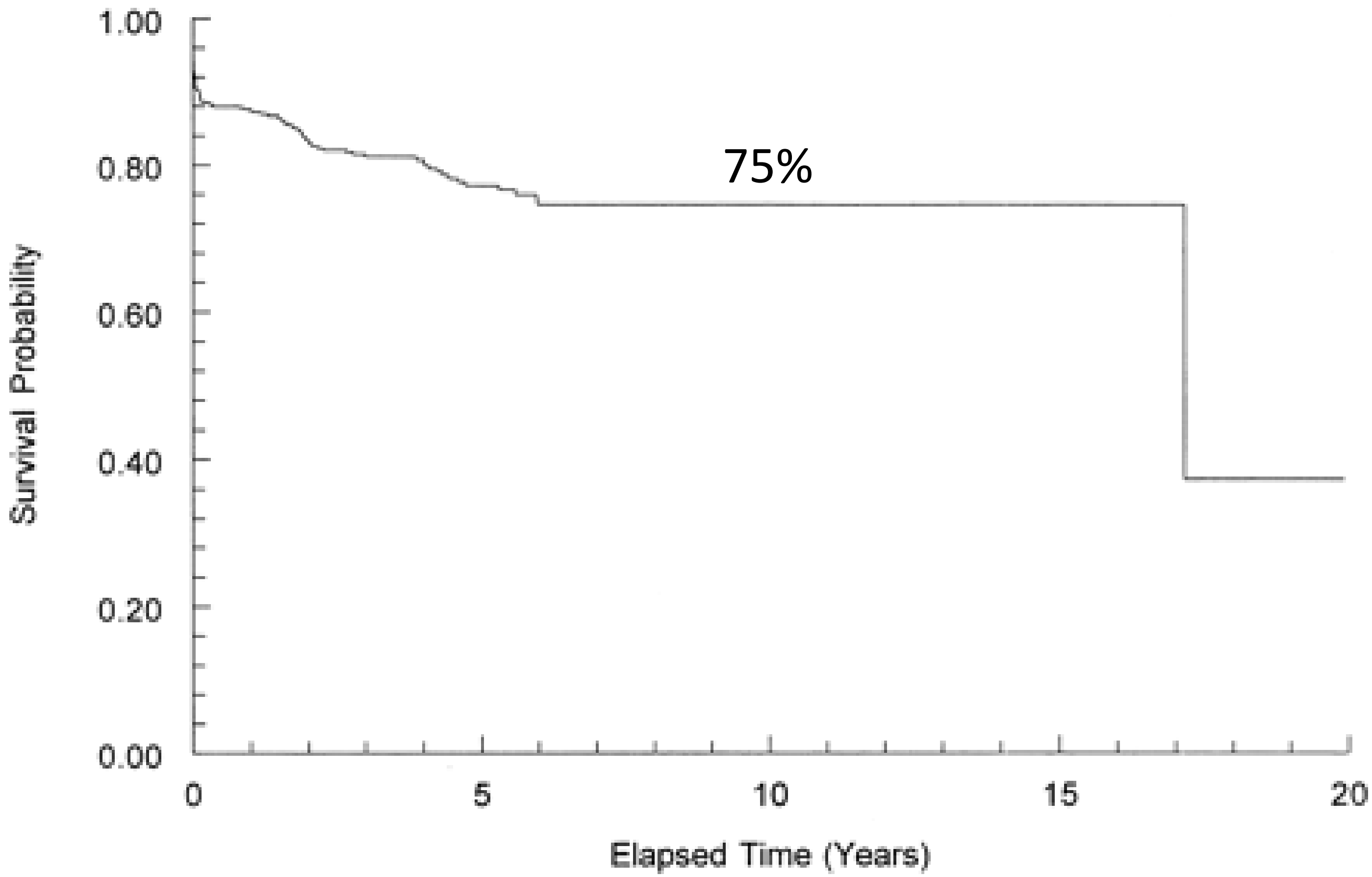


Before

12 month follow up

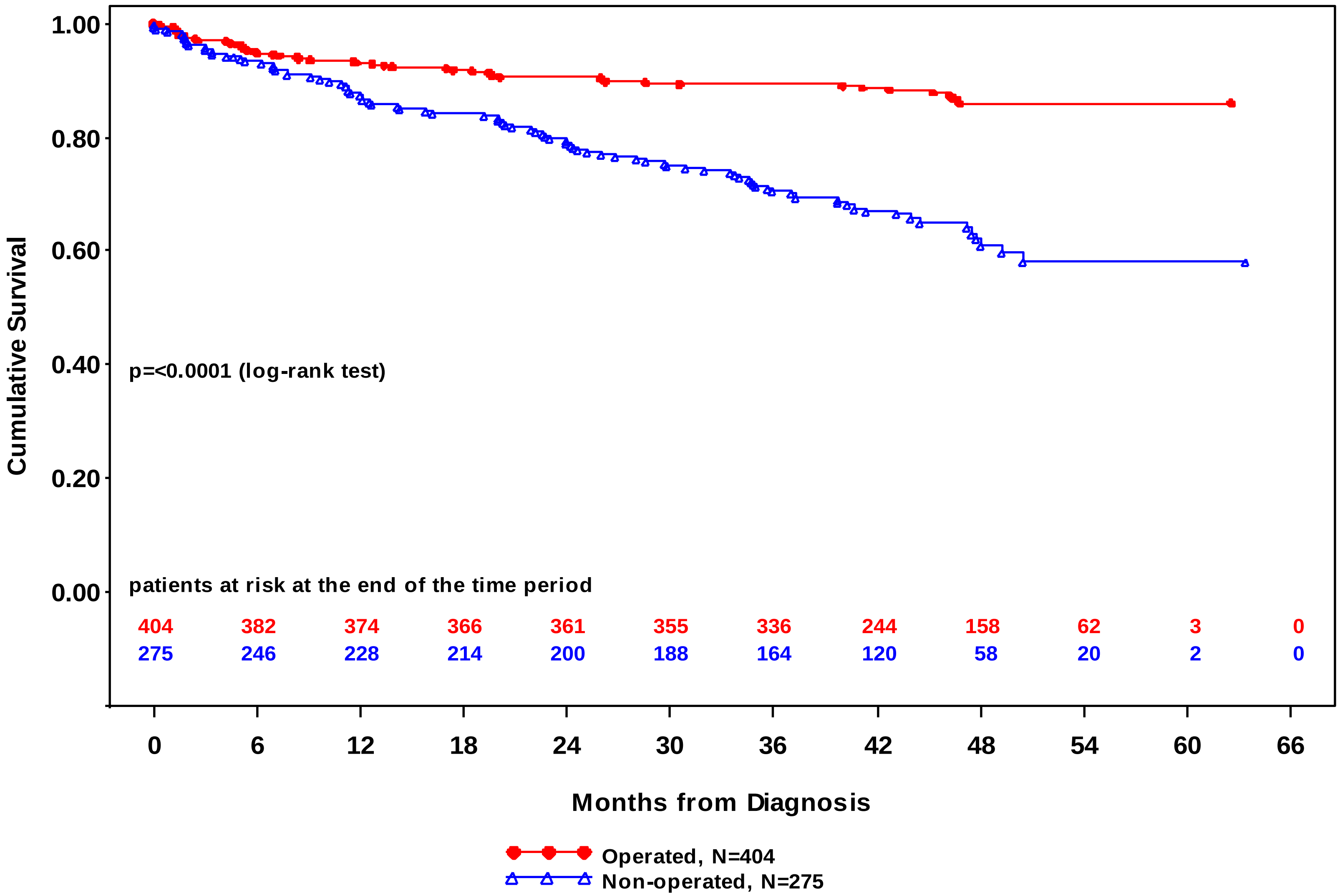
RV parameters	Pre-PEA	Post- PEA 3 months	Post- PEA 12 months	Post- PEA 24 months
TAPSE (mm)	8.8 ± 0.6	10.1 ± 0.9	11.3 ± 1.1	13.1 ± 1.4
RV-EDA (cm^2)	35.8 ± 4.4	26.6 ± 4.8	21.2 ± 5.1	20.6 ± 5.1*
RV-ESA (cm^2)	27.1 ± 3.8	17.9 ± 3.8	13.3 ± 4.1	12.5 ± 4.2*
RV-FAC (%)	24.5 ± 3.7	33.5 ± 4.62	38.4 ± 4.9	40.0 ± 4.7*
RV-MPI	0.8 ± 0.1	0.5 ± 0.1	0.4 ± 0.1	0.4 ± 0.1*

Survie à long terme après endartériectomie pulmonaire



AJRCCM 1999;160:523

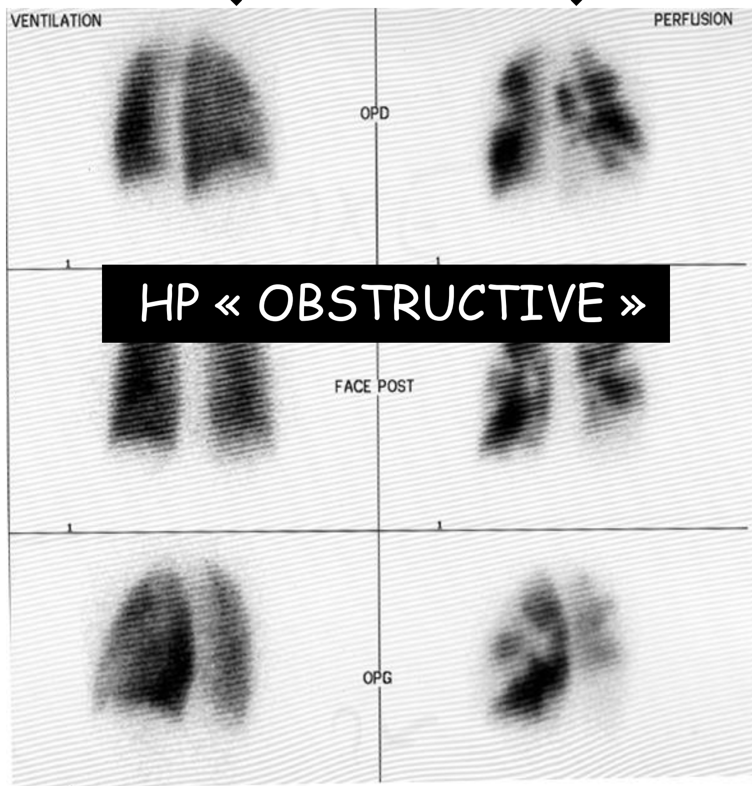
Survie Cohorte Européenne



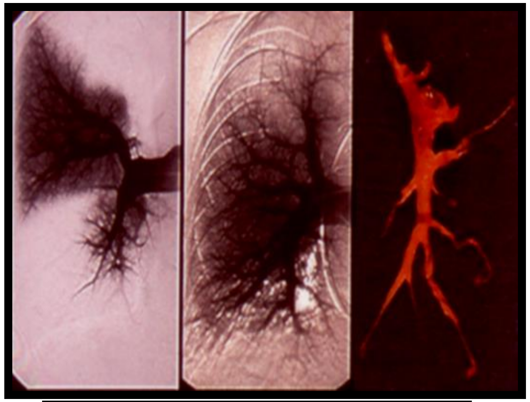
International CTEPH registry

Diagnostics différentiels

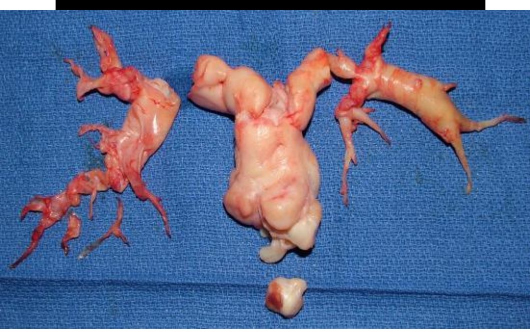
HP « OBSTRUCTIVE »



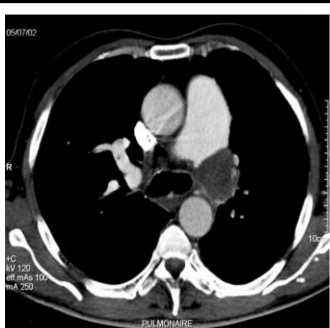
HTAP ++++



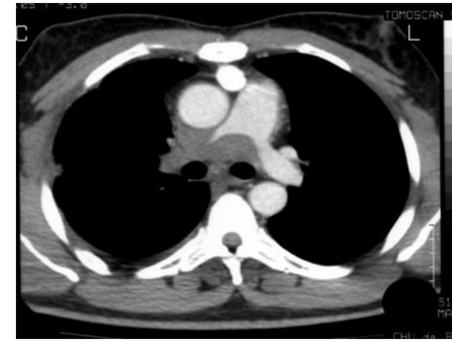
Angiosarcome pulmonaire



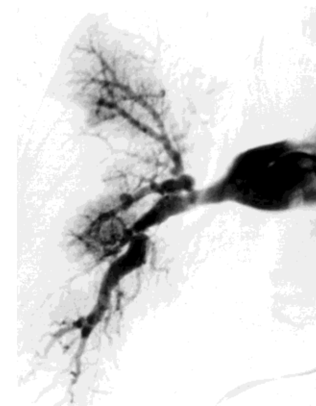
Kyste hydatique



Médiastinite fibreuse



Artérite pulmonaire



Leiomyome

Transplantation pulmonaire

Lever le barrage pulmonaire

Cependant:

-Persistance des anomalies VD

-Dysfonction diastolique du VG

-Oedème de reperfusion

-Dysfonctions associées possibles : rein, foie,…

Capacité du coeur à compenser immédiatement l'hémodynamique post-transplantation?

Journal of the American College of Cardiology

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Electrophysiologic Remodeling of the Left Ventricle in Pressure Overload-Induced Right Ventricular Failure

Maxim Hardziyenka, MD, PhD,*† Maria E. Campian, MD,* Arie O. Verkerk, PhD,*
Sulaiman Surie, MD,‡ Antoni C. G. van Ginneken, PhD,* Sara Hakim, BSc,§
André C. Linnenbank, PhD,* H.A.C.M. Rianne de Bruin-Bon, BSc,|| Leander Beekman, BSc,*
Mart N. van der Plas, MSc,‡ Carol A. Remme, MD, PhD,* Toon A. B. van Veen, PhD,§
Paul Bresser, MD, PhD,*‡ Jacques M. T. de Bakker, PhD,*†§ Hanno L. Tan, MD, PhD*||

Amsterdam and Utrecht, the Netherlands

Conclusions

Electrophysiologic remodeling occurs in the atrophic LV of CTEPH patients with RVF and in the MCT rat model of PAH-induced RVF. This is associated with abnormal LV diastolic filling.

Remodelage VD chez les patients HP

Dysfonction VD Post-transplantation

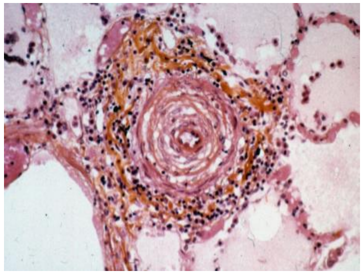
- Hypertrophie non adaptée
- Dilatation
- Septum paradoxal
- Fonctions diastolique and systolique VD altérée
- Fonction diastolique VG altérée

TxP

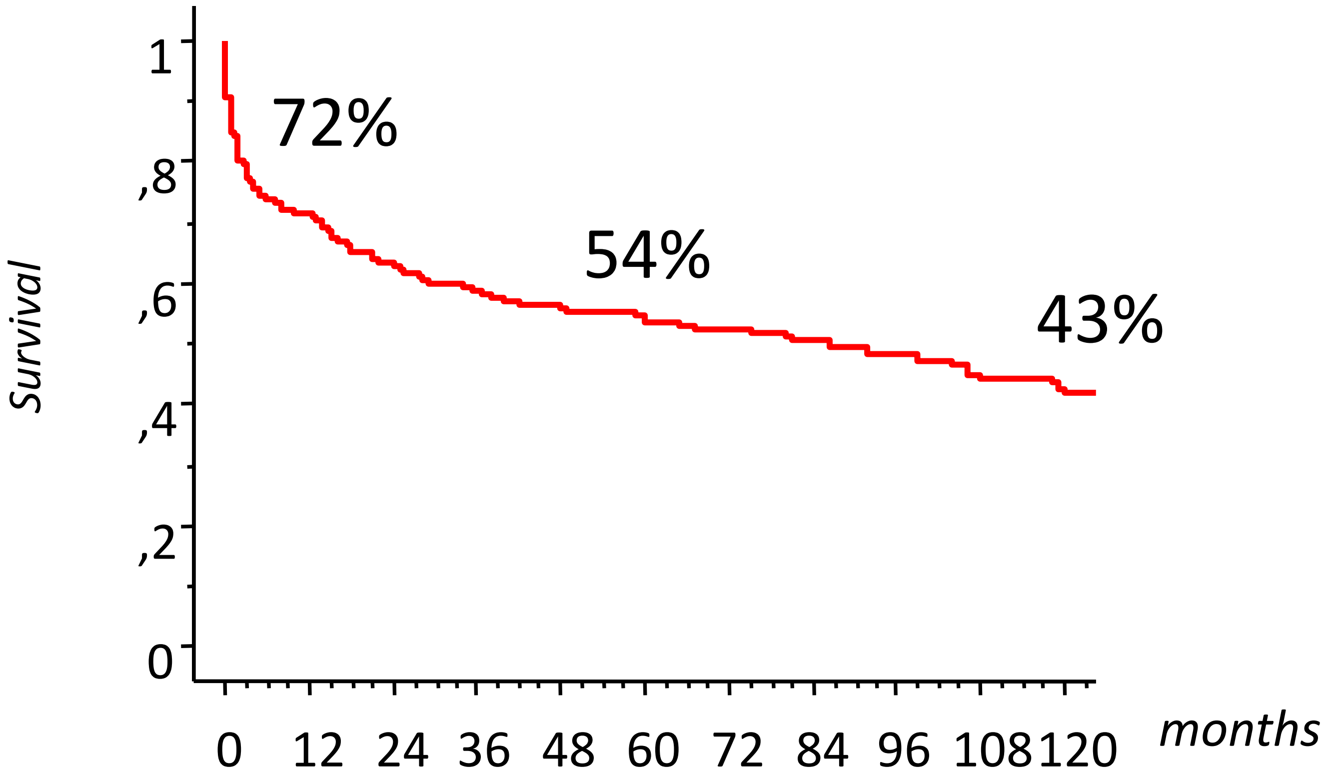
- Amélioration fonction VD
- Dysfonction diastolique VG démasquée

Oedème pulmonaire

Diminution drastique de la post-charge du VD



Marie Lannelongue Experience with PH
1986-2013
n=295

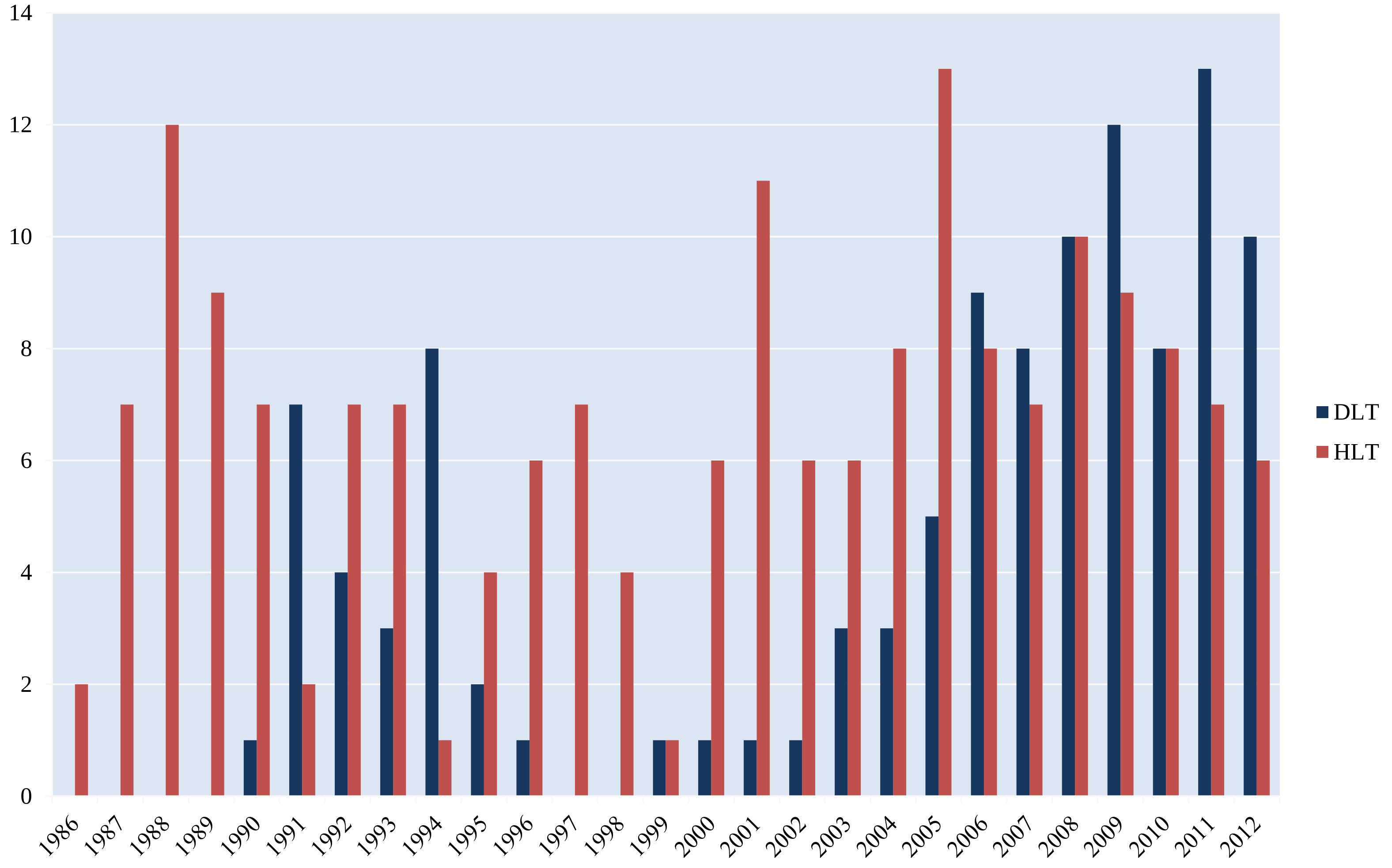


Pts: 295 202 108 54

Transplantation for PH 1986-2013

n=295

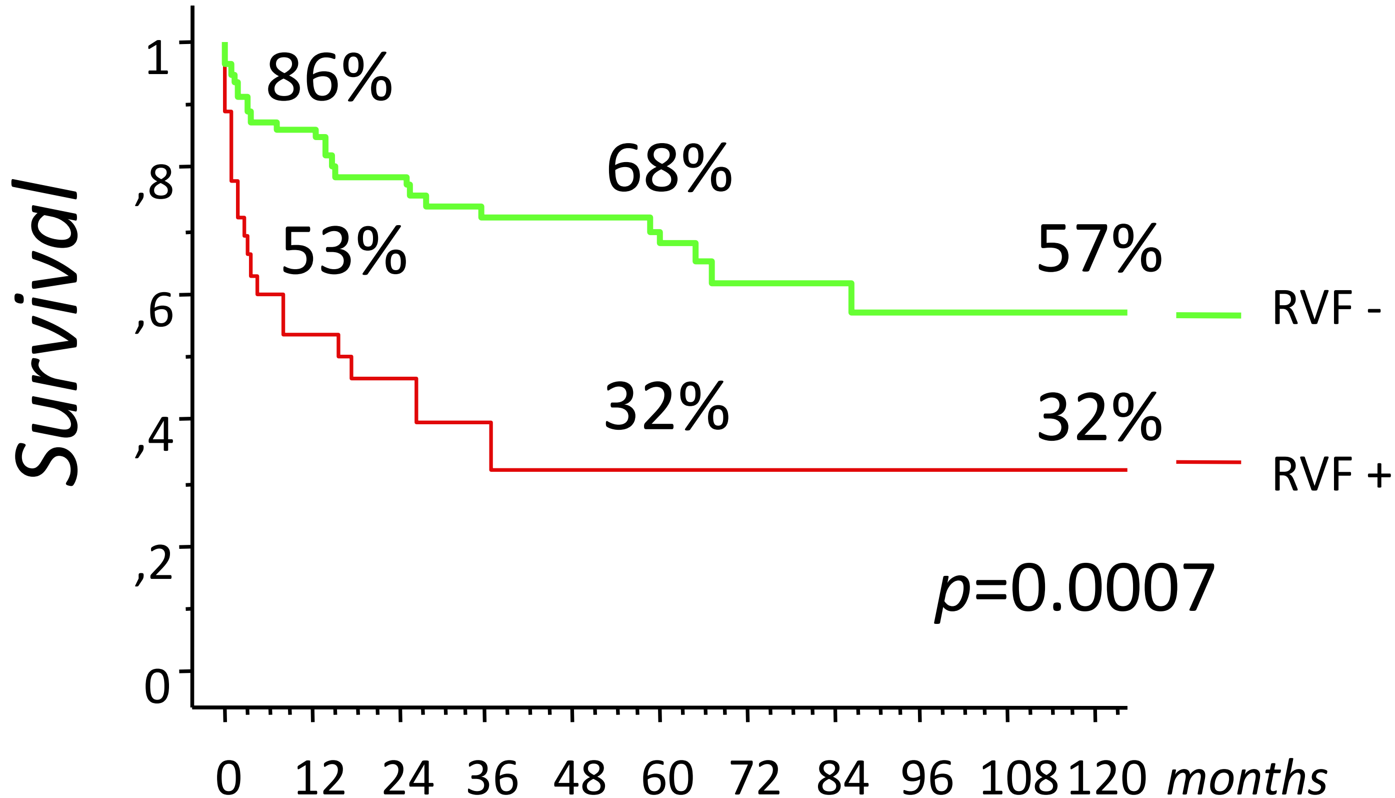
DLT=116
HLT=179



Double lung transplantation

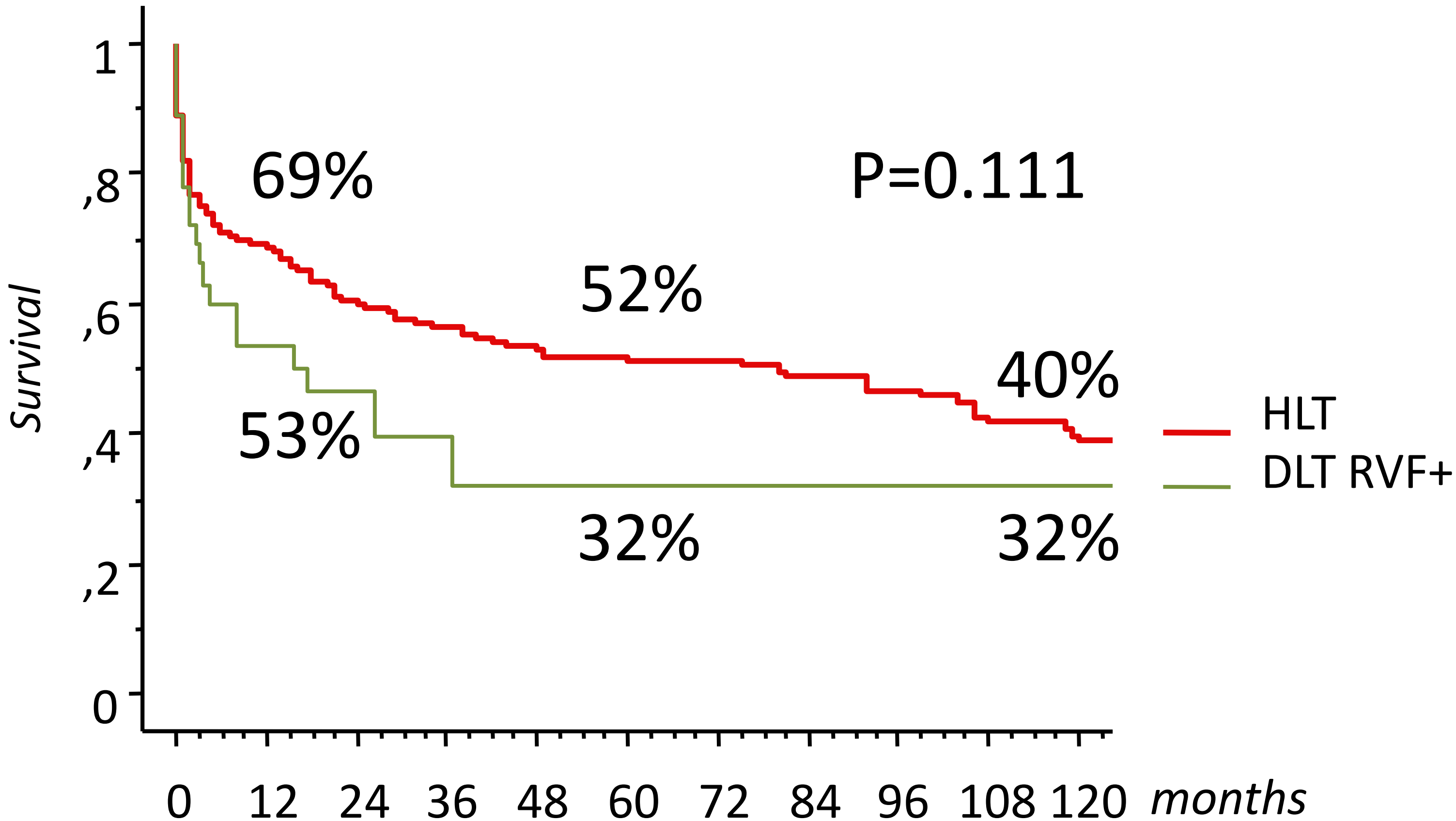
n=116

RVF | Inotropic support
Syncope within
the month



RHF +:	36	17	4	2
RHF -:	80	62	31	12

HLT vs. DLT RVF+

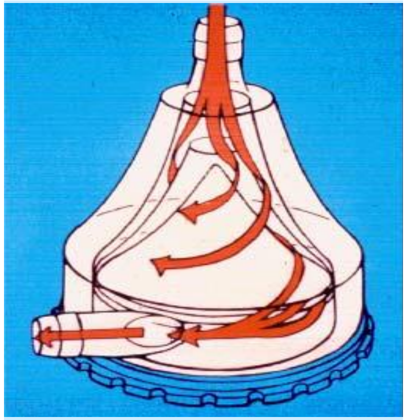


RHF +:	36	17	4	2
HLT:	179	123	73	40

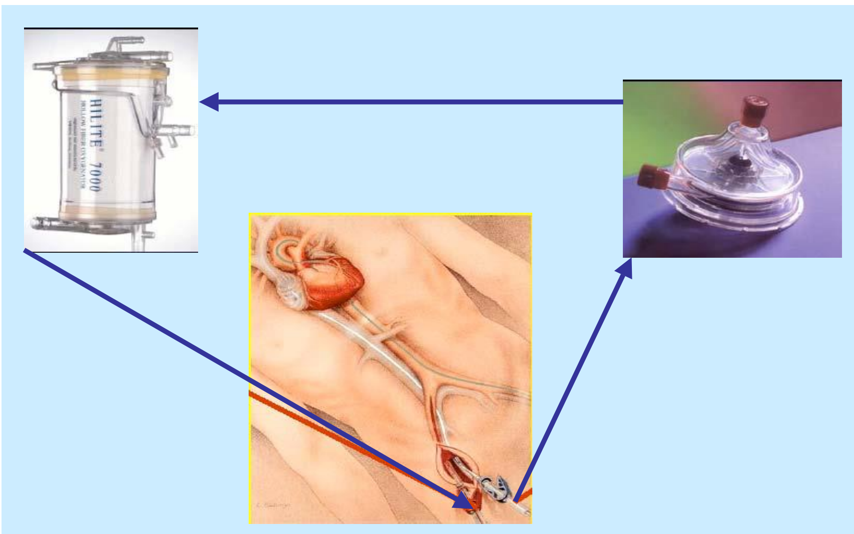
HLT | ECLS 12
ES 36

Avancées techniques

- Oxygénateurs en polymethylpentène
- Tubulures héparinées
- Pompes centrifuges



Terminologies ECLS
ExtraCorporeal Life Support



ECMO = Extracorporeal Membrane Oxygenation

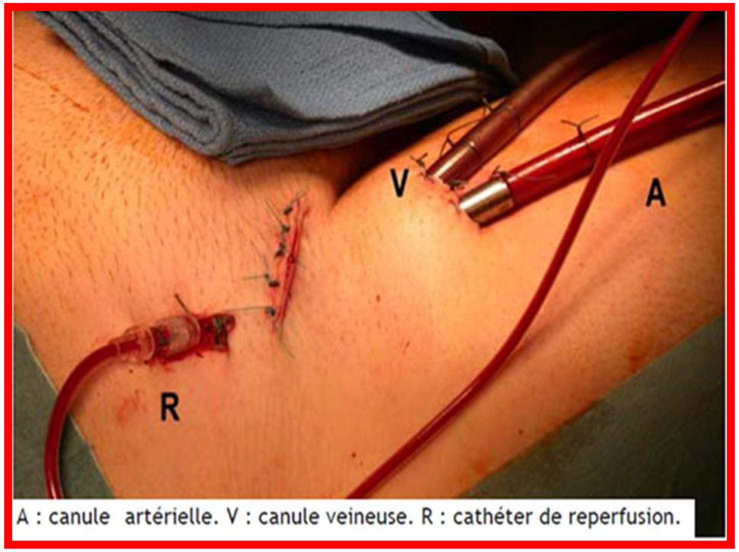
Decarboxylation = ECMO low-flow, PECOR (Partial Extracorporeal Carbone Dioxyde Removal), ECCO^{2}R (Extracorporeal CO^2 Removal)

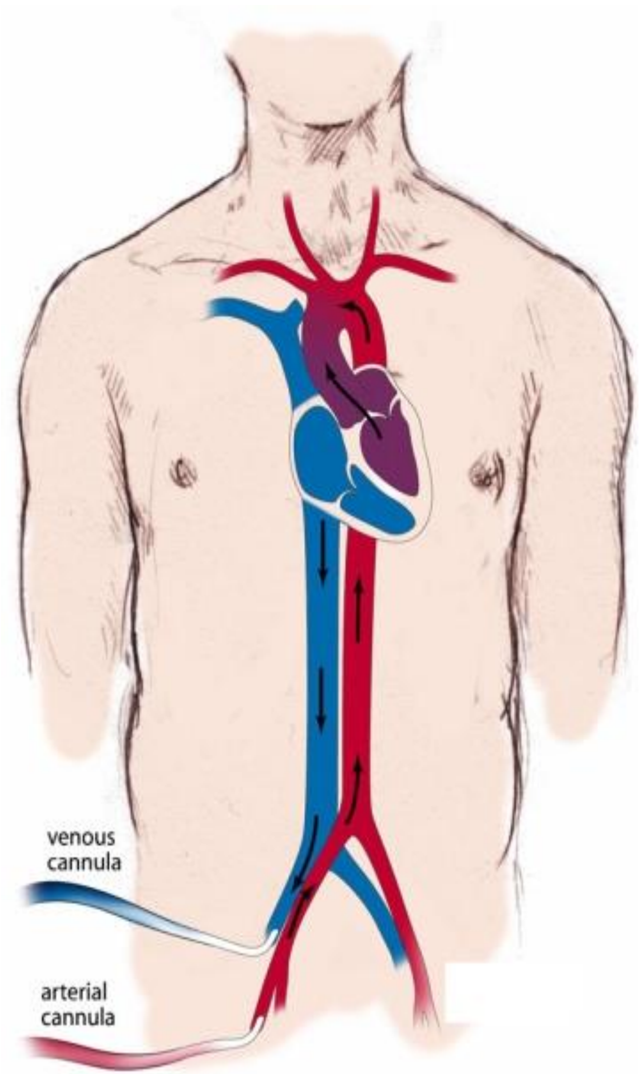
Oxygénation et decarboxylation = ECLA (ExtraCorporeal Lung Assist)

V-V, V-A, V-V-A **ECMO**

Accès Vasculaire

V-A ECMO



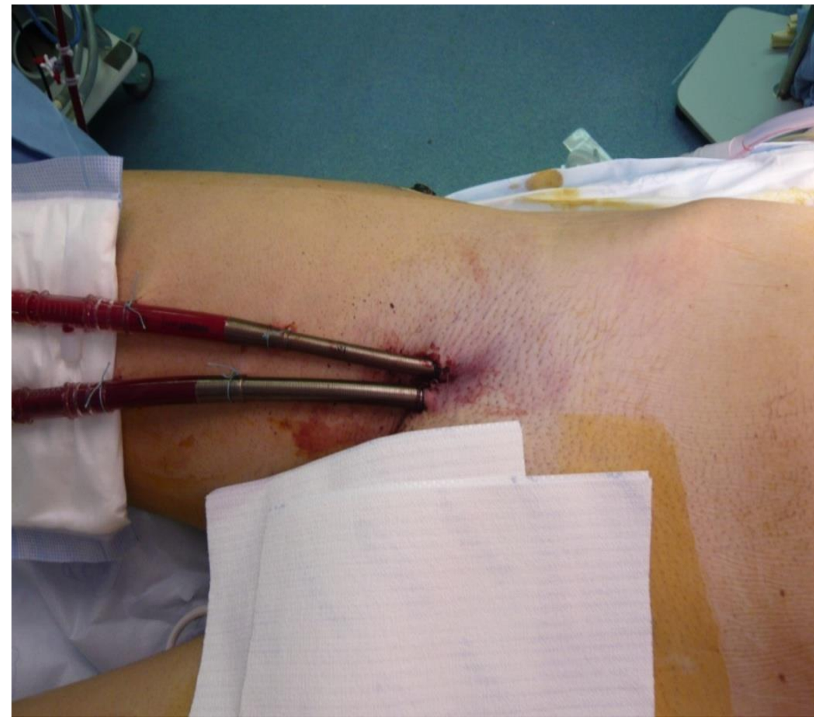


- Rapidité ++ Anesthésie locale
- Bridge +++
- Obligation de réinjection FS
- Pb de flux compétitif
- Pb de décharge G
- Infection du scarpa

ECLS pour récupération du ventricule droit?

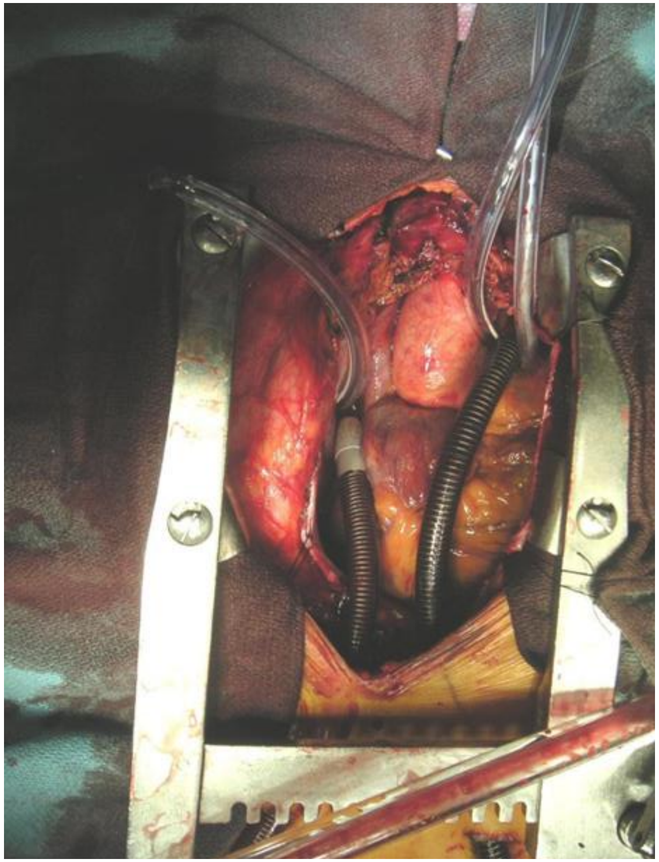
Novalung Central

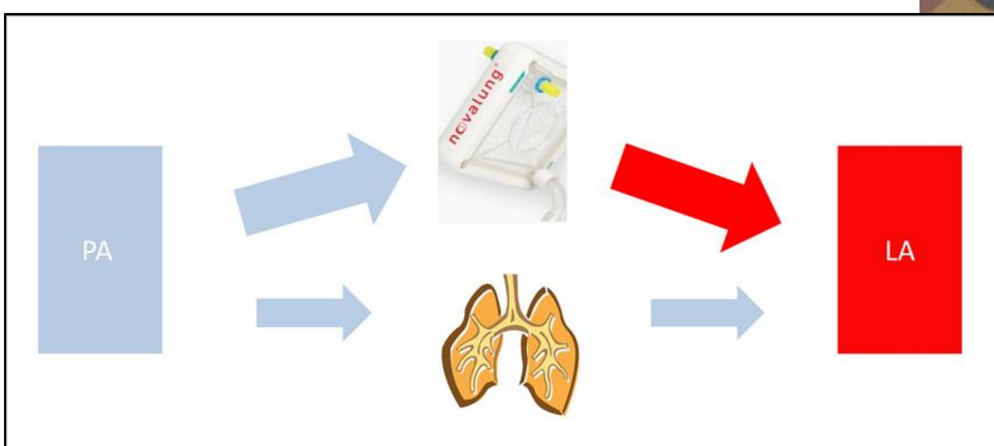
VA ECMO



Assistance de courte durée => en pont à la transplantation ou à la récupération

PA-LA Novalung*





De Perrot, JHLT 2011

Table 1 Patient Characteristics at the Time of Listing

Variable[a]	1998–2005 (n = 23)	2006–2010 (n = 21)	p-value
Age, years	35 ± 15	37 ± 18	0.7
Female sex	18 (78)	14 (67)	0.7
NYHA class	3.1 ± 0.3	3.4 ± 0.6	0.04
6-MWT distance, meters	343 ± 92	293 ± 127	0.1
Right atrial pressure, mm Hg	14 ± 8	11 ± 6	0.2
Cardiac output, liters/min	3.3 ± 0.7	3.7 ± 0.9	0.1
PVR, dynes · sec · cm^{-5}	1173 ± 314	1145 ± 537	0.8
Patients on IV epoprostenol	18 (78)	13 (62)	0.2
Duration of IV epoprostenol, mon	16 ± 17	21 ± 27	0.5
Patients on oral therapy	8 (35)	14 (67)	0.002

IV, intravenous; NYHA, New York Heart Association; PVR, pulmonary vascular resistance.
[a]Data are presented as number (%) or mean ± standard deviation.

Table 2 Pre-transplant Management

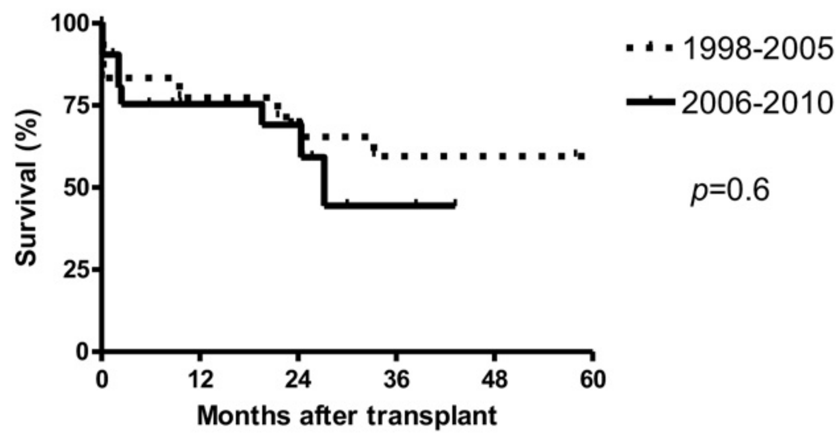
Variable	1998–2005 (n = 23) No. (%)	2006–2010 (n = 21) No. (%)	p-value
In-hospital pre-transplant	1 (4)	10 (48)	0.0009
Atrial septostomy	2 (9)	0	0.2
Extracorporeal life support	0	6 (29)	0.006
PA-LA Novalung	0	4	
VA ECMO	0	2	
Inotropic support	0	5 (25)	0.01
Intubated	0	4 (20)	0.02
Waiting list mortality	5 (22)	0	0.03
Type of transplant			0.05
Bilateral lung	18	17	
Heart-lung	0	4	

ECMO, extracorporal membrane oxygenation; LA, left atrium; PA, pulmonary artery; VA, venoarterial.

Table 4 Early Post-transplant Outcome

Variable	1998–2005 (n = 18)	2006–2010 (n = 21)	p-value
30-day mortality, No.	3	2	0.5
Severe PGD[a], No.	4	5	0.9
LOS (mean ± SD)			
Intensive care unit	17 ± 13	36 ± 30	0.02
Hospital	35 ± 27	66 ± 68	0.08

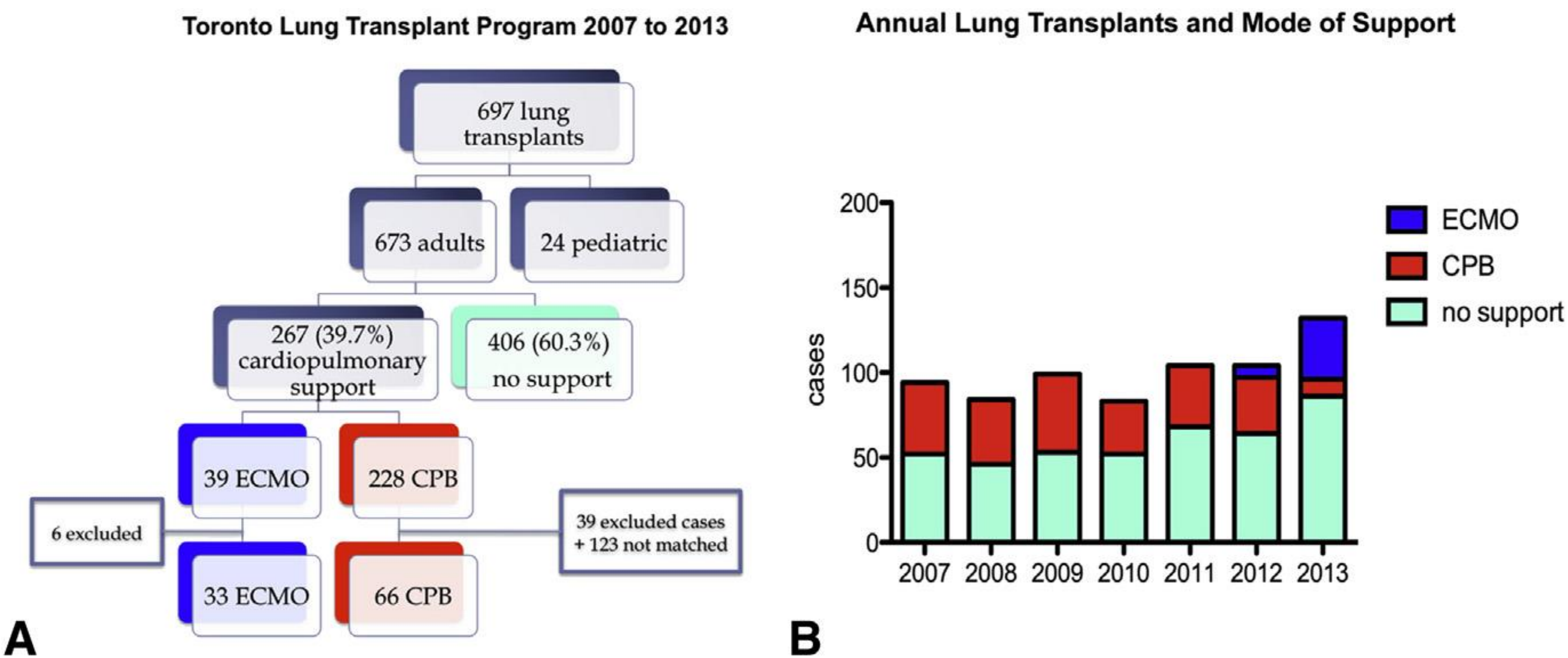
LOS, length of stay; PGD, primary graft dysfunction; SD, standard deviation.
[a]Defined by PGD III persistent during the initial 72 hours after transplant.



De Perrot, JHLT 2011

Outcomes of intraoperative extracorporeal membrane oxygenation versus cardiopulmonary bypass for lung transplantation

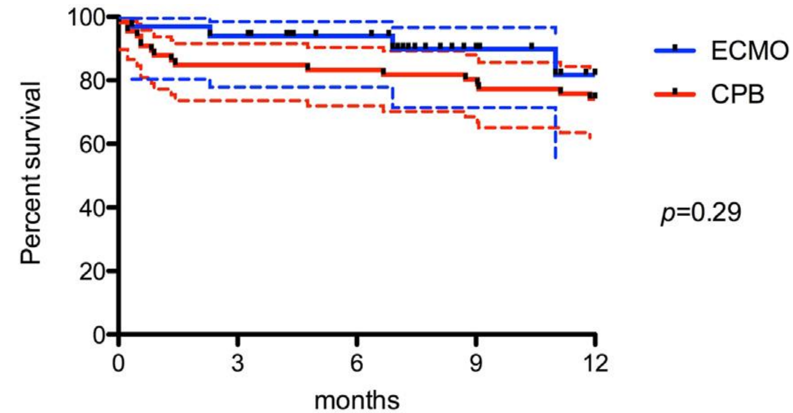
Tiago N. Machuca, MD, Stephane Collaud, MD, MSc, Olaf Mercier, MD, PhD, Maureen Cheung, MD, Valerie Cunningham, CCP, S. Joseph Kim, MD, PhD, Sassan Azad, CRA, Lianne Singer, MD, MSc, Kazuhiro Yasufuku, MD, PhD, Marc de Perrot, MD, MSc, Andrew Pierre, MD, MSc, Karen McRae, MD, Thomas K. Waddell, MD, PhD, Shaf Keshavjee, MD, MSc, and Marcelo Cypel, MD, MSc



JTCVS 2015

TABLE 2. Intraoperative data and early post-transplant outcomes

Variable	CPB (n = 66)	ECMO (n = 33)	*P* value
Intraoperative			
Pump time (min)	199 ± 90	210 ± 84	.38
Warm ischemic time L (min)	69 ± 17	73 ± 23	.37
Warm ischemic time R (min)	64 ± 14	70 ± 20	.123
Warm ischemic time SLTx (min)	74 ± 9	74 ± 19	.94
Blood transfusion			
pRBCs (units)	6 (3-10)	3 (2-5.5)	<.001
Platelets (units)	1 (0-2)	0 (0-0.5)	.007
FFP (units)	4 (1.7-7.2)	0 (0-0.5)	.006
Postoperative			
pRBC up to 72 h (units)	2 (1-6)	1 (0.5-3)	.014
Length of MV (d)	7.5 (2-18)	3 (2.5-5)	.005
ICU stay (d)	9.5 (3-21)	5 (3-9)	.026
Hospital stay	27 (17-42)	19 (14-30)	.029
ECLS postoperative requirement	5 (7.5%)	0	.166
Dialysis requirement	12 (18%)	3 (9%)	.37
Reoperation (bleeding)	18 (27%)	3 (9%)	.04
90-d mortality	10 (15%)	2 (6%)	.32



Patients at risk:

ECMO	33	32	26	14	8
CPB	66	57	56	54	49

JTCVS 2015

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Institutional experience with extracorporeal membrane oxygenation in lung transplantation§

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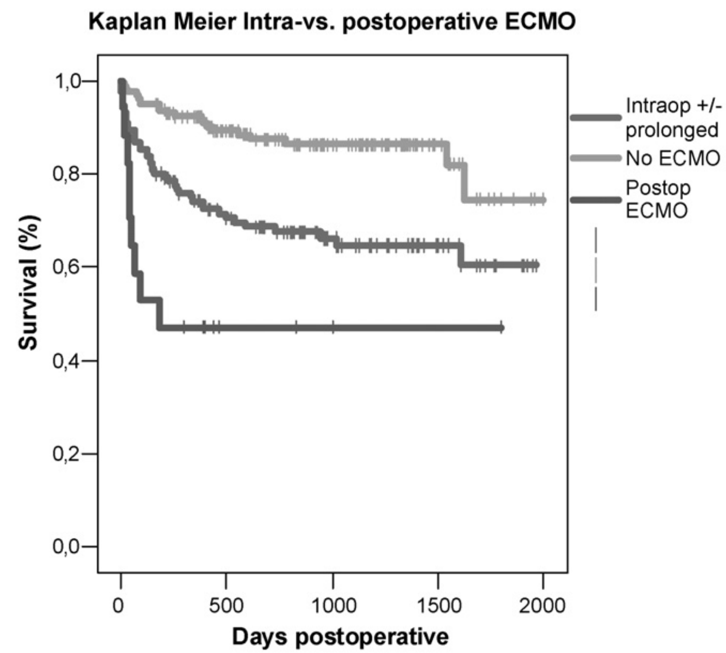
Table 3
Overview of ECMO and CPB use

	Bridge to TX	Intraoperative	Prolonged	Postoperative
ECMO	2	130	51	5
CPB±ECMO		27	0	11
No support		149	0	6

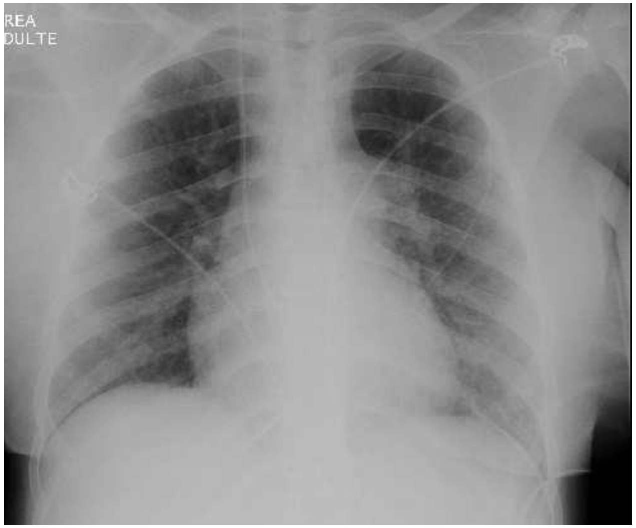


p (Intraop vs. no) < 0,01
p (Postop vs. no) < 0,01
p (Intraop vs. Postop.) = 0,02

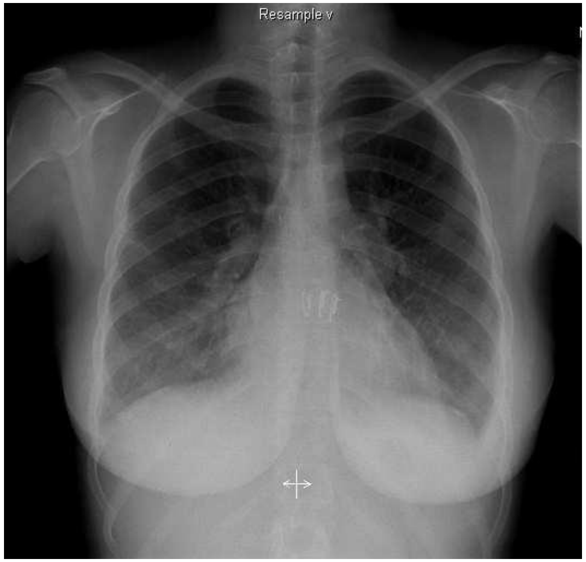
Patients at risk: 3 months n= 257
1 year n= 208
3 years n= 100

Innovation Prise en charge post-Tx pulmonaire des patients PH

- **Apports du KT OG**
  - Monitorage du remplissage
  - Indication objective d'ECMO
  - Amélioration des suites immédiates

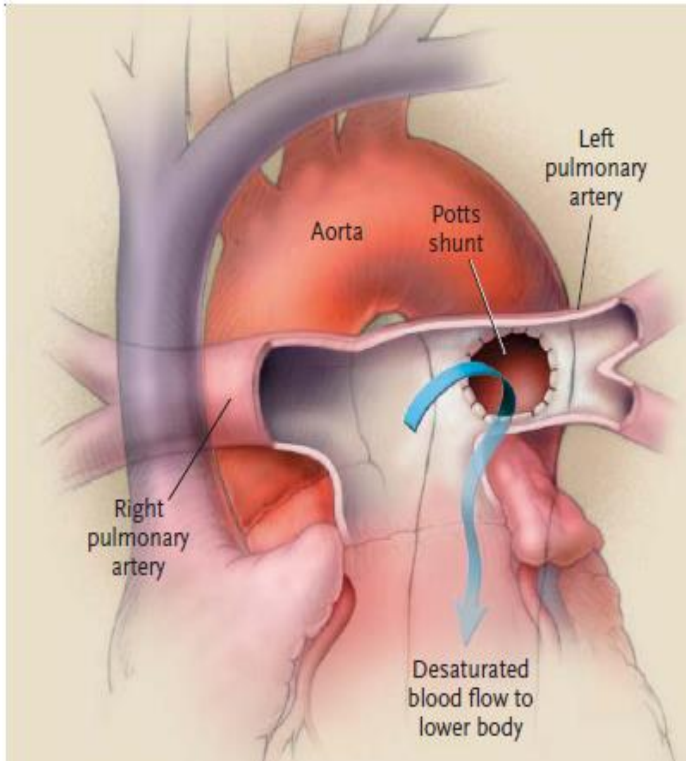


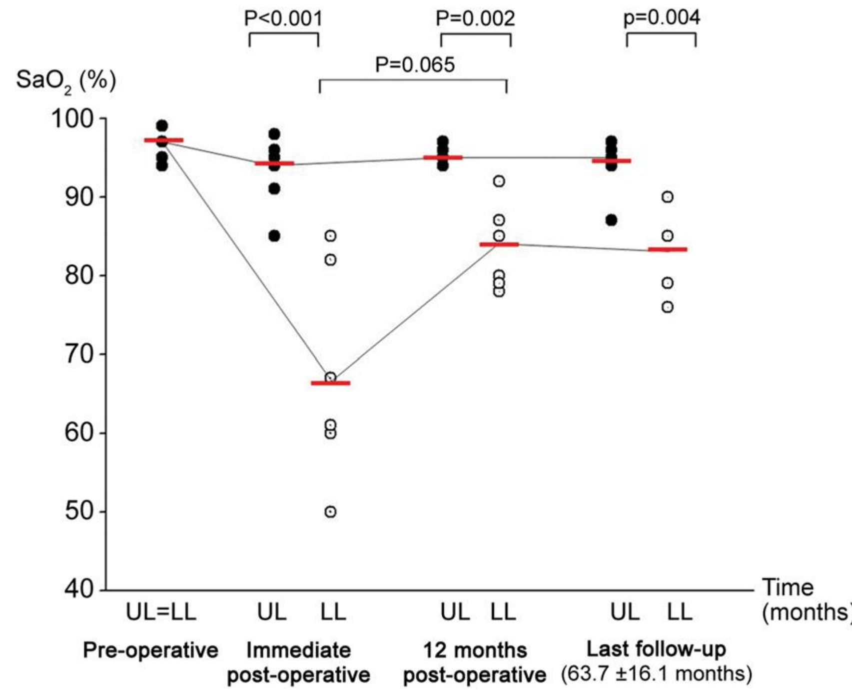
PAPm 97 mm Hg

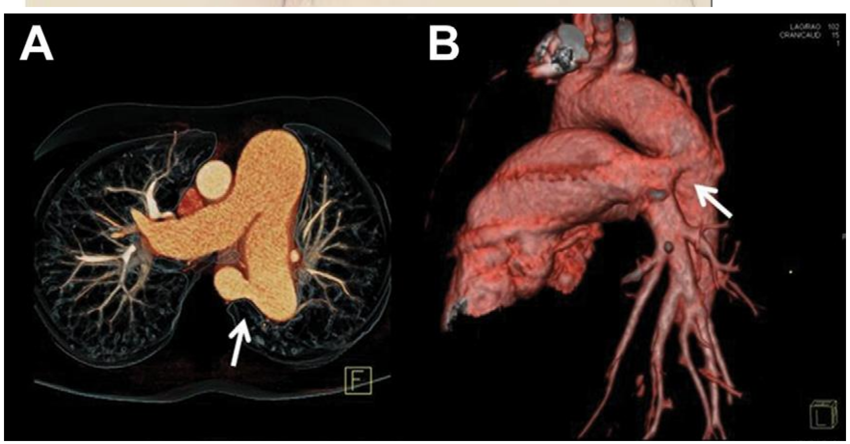


3 sem.

Potts Procedure In Children

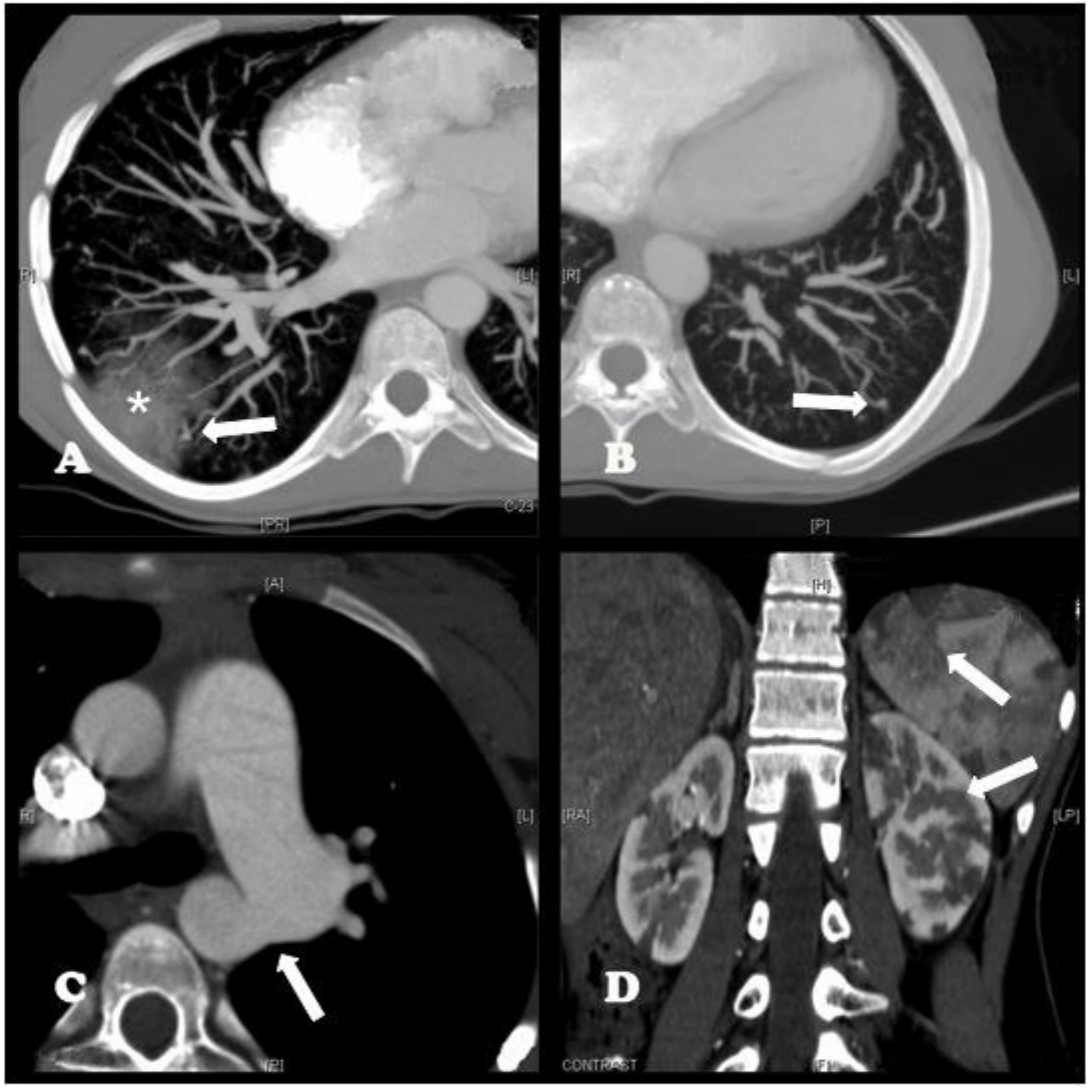






- Supra-systemic PH in Children
- Improvement in NYHA functional class
- Delay Tx (5-6y)
- Carries acceptable risk

Baruteau, ATS 2012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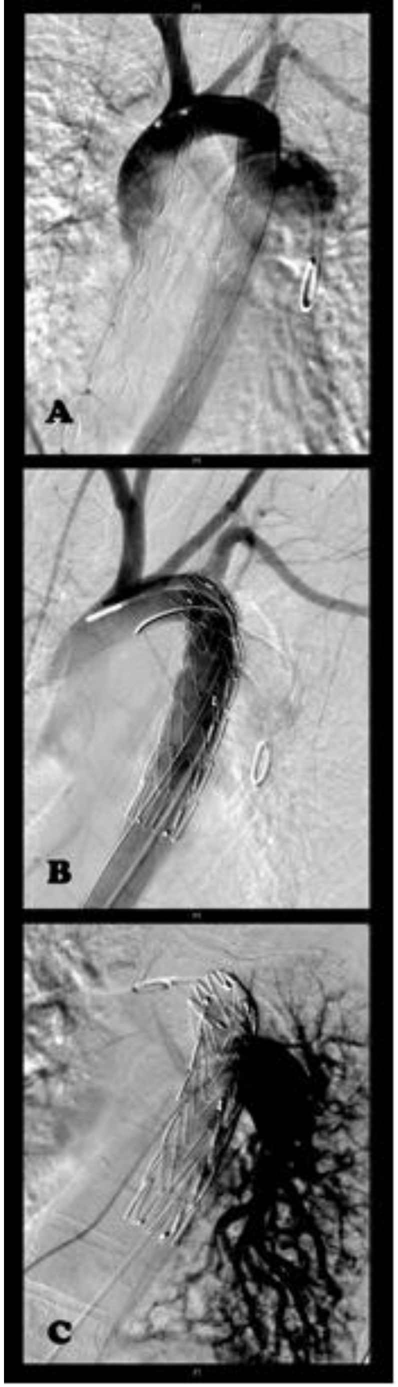
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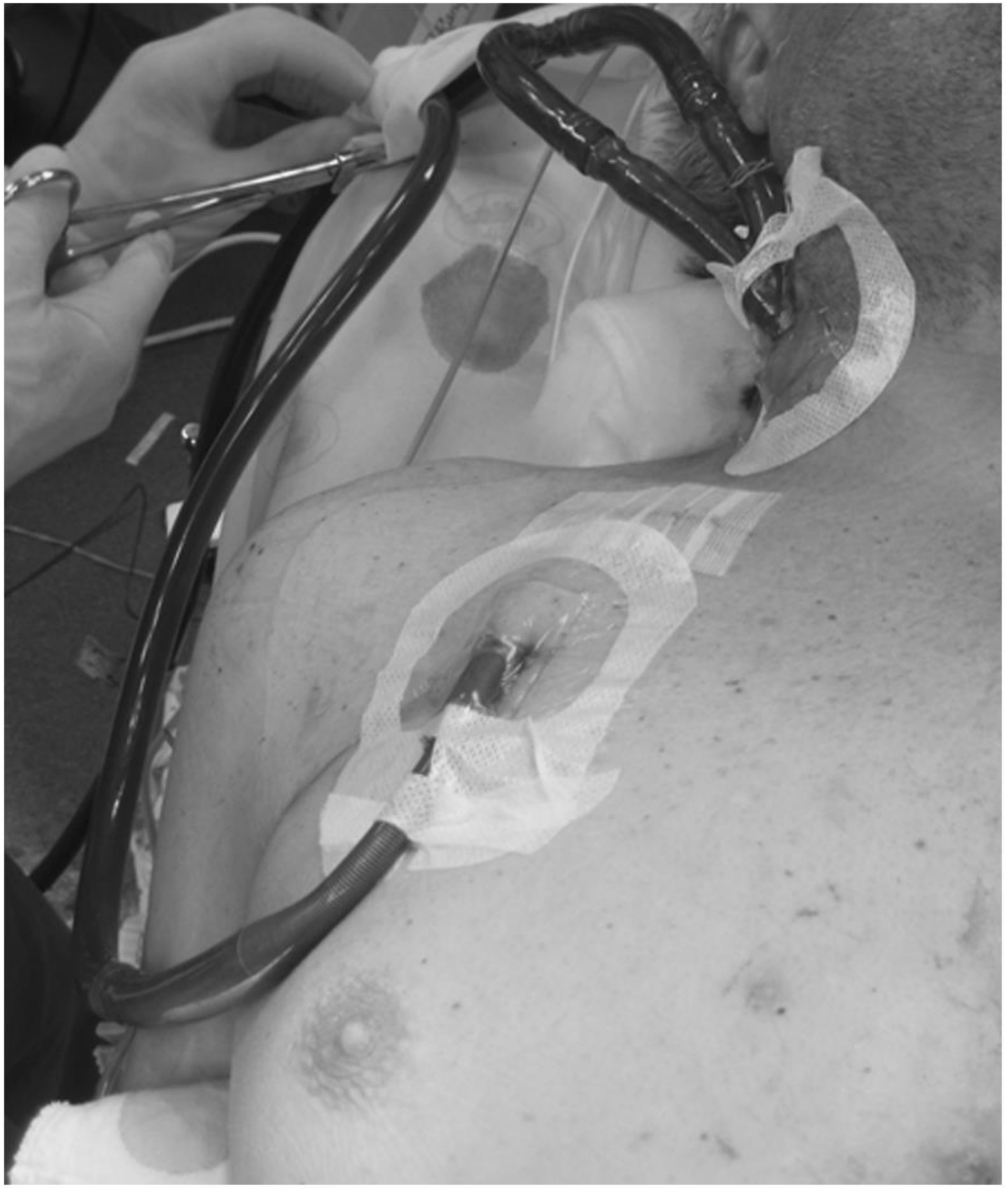
B
C



Javidfar, Curr Opin Organ Transplant 2012

Bio-Artificial Lung
for the treatment of
Pulmonary Hypertension Induced
End-stage Right Ventricle Failure

BIOART-LUNG 2020

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