Transplantation Pulmonaire

Pour hypertension pulmonaire

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Service de Transplantation Pulmonaire

Hôpital Marie Lannelongue

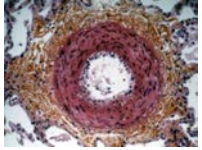
Vendredi 23 Juin 2017 – Cours DES Pneumologie

Transplantation pulmonaire

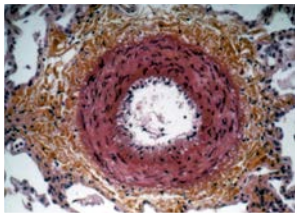
Pour hypertension pulmonaire

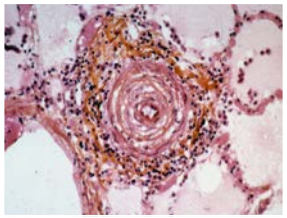
- Introduction
- Référer et inscrire
- Type de procédure TCP vs TBP
- Impact de la dysfonction VD
- HTAP associée aux cardiopathies congénitales
- Bénéfices de la superU
- Transplantation pour SSc

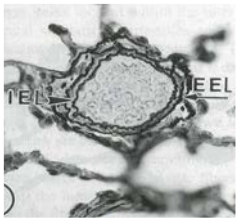
Introduction

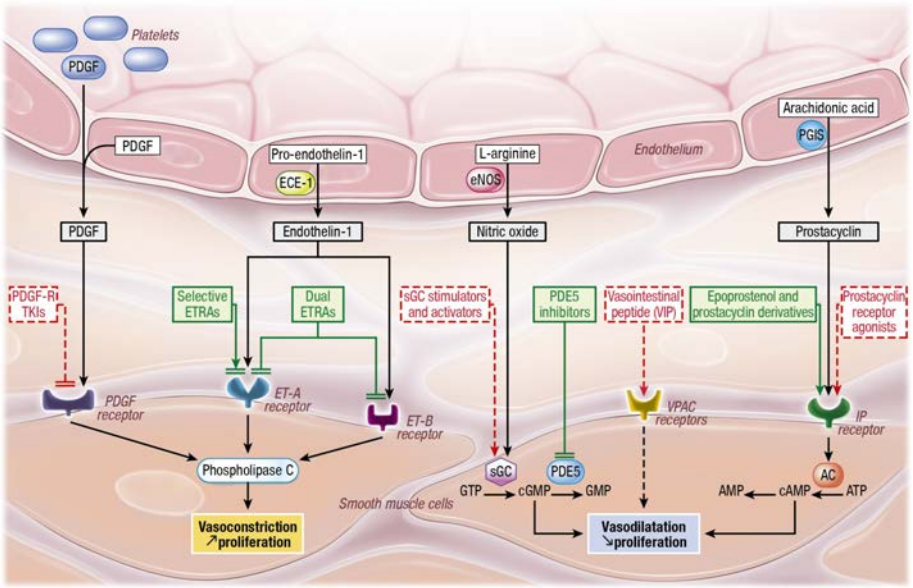


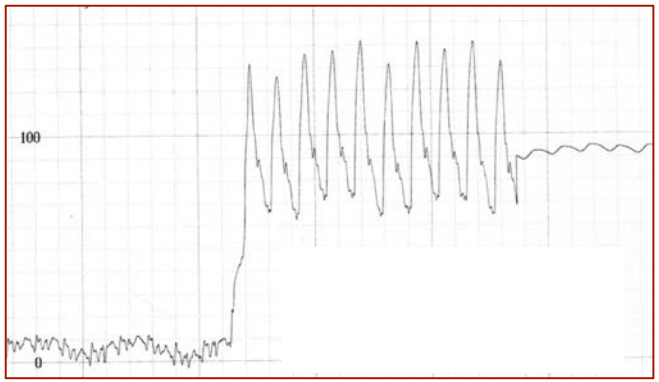
Transplantation pulmonaire
Hypertension artérielle pulmonaire

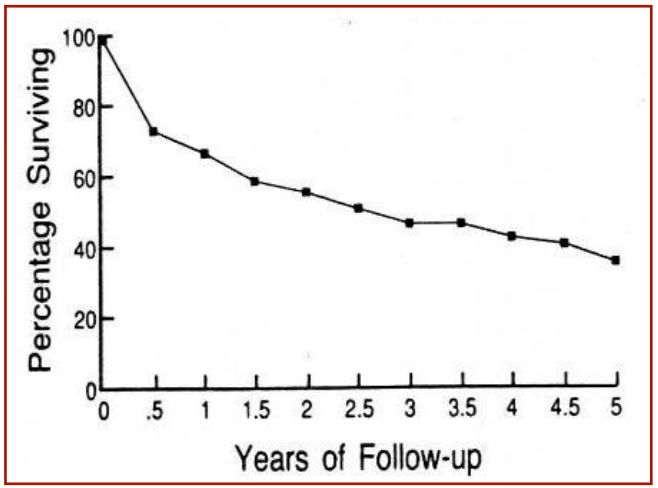










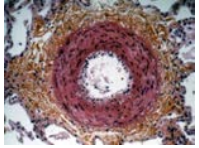


O'Callaghan DS et al. *Nat Clin Practice Cardiol* 2011

D'Alonzo GE et al, *Ann Int Med* 1991

Classification

Hypertension artérielle pulmonaire



1. Pulmonary Arterial Hypertension (PAH)

- 1.1. Idiopathic
- 1.2. Heritable
  - 1.2.1. BMPR2
  - 1.2.2. ALK1, endogline (with or without HHT)
  - 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 diseases
  - 1.4.5. Schistosomiasis
  - 1.4.6. Chronic haemolytic anemias
- 1.5. Persistent PH of the newborn

1'. PVOD & PCH

2. PH due to left heart disease

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

3. PH due to lung diseases and/or hypoxia

- 3.1. COPD
- 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

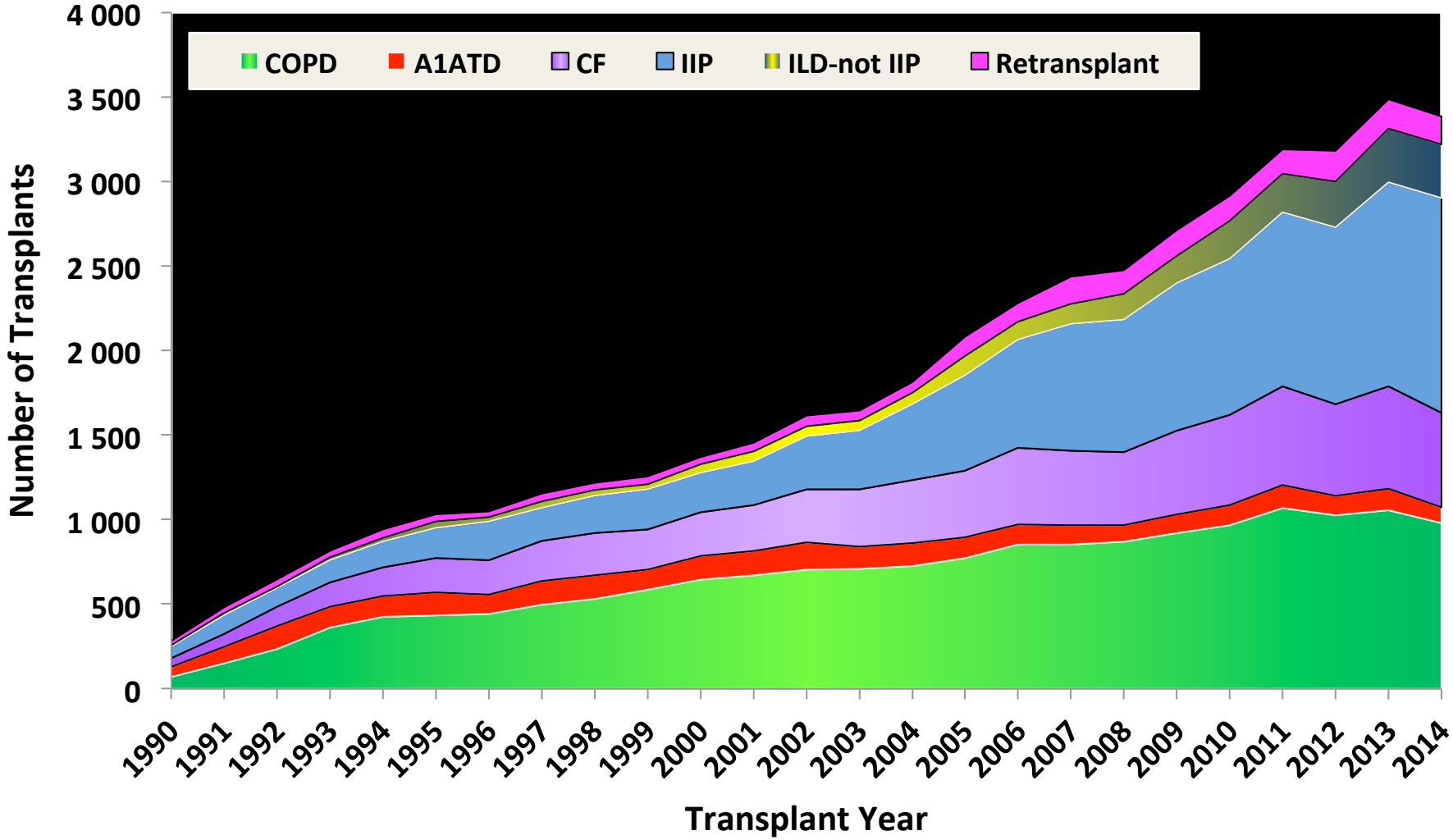
4. Chronic thromboembolic PH (CTEPH)

5. PH with unclear and/or multifactorial mechanisms

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

Simonneau G, et al. *JACC* 2009

Adult Lung Transplants
Major Indications by Year (Number)



ISHLT • INTERNATIONAL SOCIETY FOR HEART AND LUNG TRANSPLANTATION

JHLT. 2016 Oct; 35(10): 1149-1205

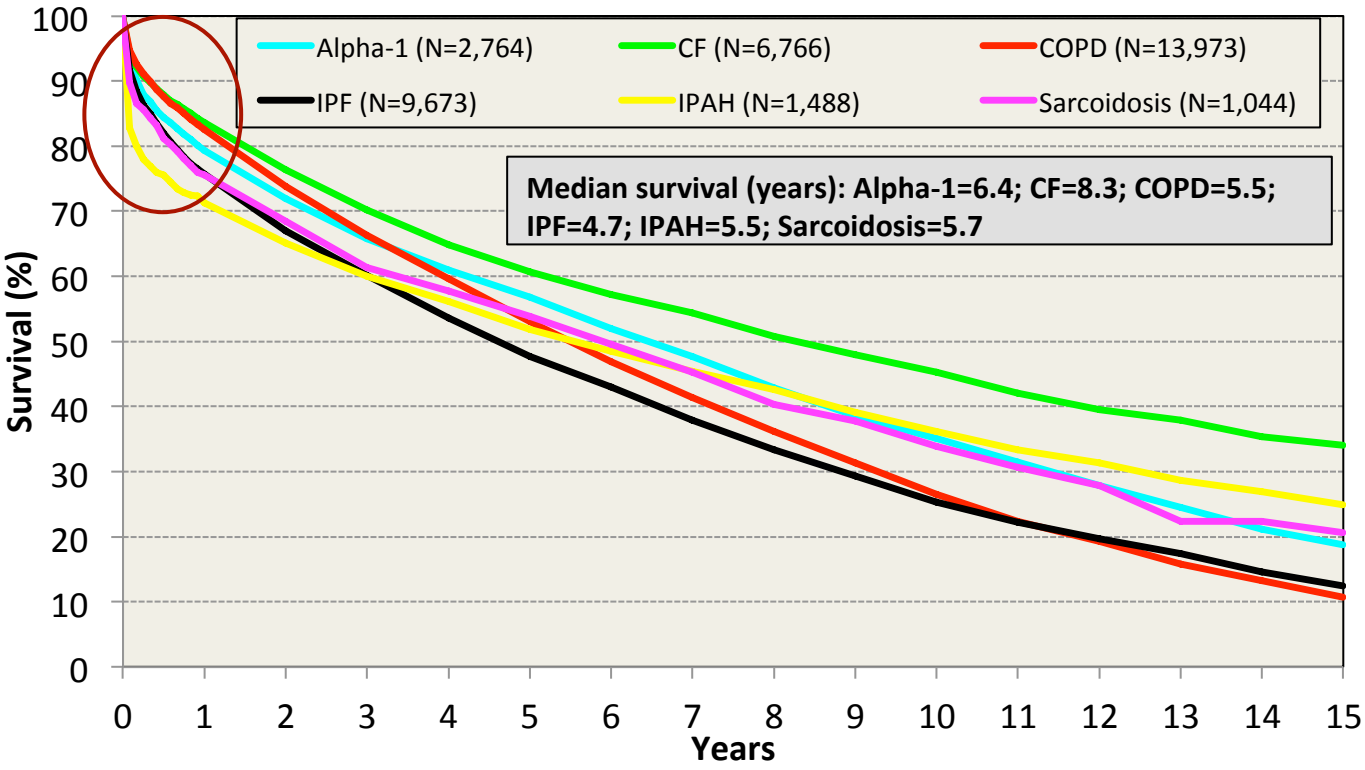
Adult Lung Transplants

(Transplants: January 1995 – June 2015)

Diagnosis	SLT (N=17,213)	BLT (N=32,789)	TOTAL (N=50,002)
COPD	6,999 (40.7%)	8,674 (26.5%)	15,673 (31.3%)
IIP	5,979 (34.7%)	6,264 (19.1%)	12,243 (24.5%)
CF	209 (1.2%)	7,686 (23.4%)	7,895 (15.8%)
ILD-not IIP	977 (5.7%)	1,608 (4.9%)	2,585 (5.2%)
A1ATD	784 (4.6%)	1,784 (5.4%)	2,568 (5.1%)
Retransplant	874 (5.1%)	1,174 (3.6%)	2,048 (4.1%)
IPAH	87 (0.5%)	1,348 (4.1%)	1,435 (2.9%)
Non CF-bronchiectasis	64 (0.4%)	1,293 (3.9%)	1,357 (2.7%)
Sarcoidosis	307 (1.8%)	941 (2.9%)	1,248 (2.5%)
PH-not IPAH	129 (0.7%)	648 (2.0%)	777 (1.6%)
LAM/tuberous sclerosis	141 (0.8%)	359 (1.1%)	500 (1.0%)
OB	75 (0.4%)	354 (1.1%)	429 (0.9%)
CTD	122 (0.7%)	240 (0.7%)	362 (0.7%)
Cancer	7 (0.0%)	27 (0.1%)	34 (0.1%)
Other	459 (2.7%)	389 (1.2%)	848 (1.7%)

JHLT. 2016 Oct; 35(10): 1149-1205

Transplantation pulmonaire *pour HTAP*



- Dysfonction ventriculaire droite toujours associée
- Circulation extracorporelle systématique
- Haut risque de dysfonction primaire du greffon
- Mortalité sur liste d'attente

Référer
Inscrire sur liste

Transplantation Pulmonaire
Quand référer ?

- Référer quand survie prédite à 3 ans < 50% et/ou altération fonctionnelle NYHA III-IV malgré traitement médical maximal

- Le temps d'attente est influencé par de nombreux facteurs en particuliers taille et groupe sanguin (femme > homme grand groupe A et AB)

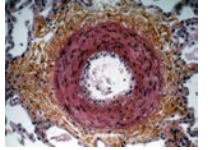
- Survie sur liste classiquement plus courte pour HTAP idiopathique, fibrose et mucoviscidose comparativement à la BPCO et au syndrome d'Eisenmenger

Weill D et al, *JHLT* 2015

	Number of Patients				
Diagnosis Group (*n*)	Died Waiting	Removed or Still Waiting	Transplanted	Mean (SD) Age*	Median (IQR) Days Waiting
Obstructive lung disease (163)	29	12	122	51 (7)	120 (45, 320)
Single lung (92)	18	6	68	53 (6)	90 (45, 295)
Double lung (35)/heart–lung	11	6	54	48 (8)	165 (38, 356)
Cystic fibrosis (174)	65	8	101	25 (7)	216 (82, 397)
Eisenmenger's syndrome (76)	15	6	55	32 (9)	552 (249, 1108)
Bronchiectasis (51)	15	10	30	46 (8)	281 (93, 661)
Pulmonary fibrosis (100)	33	7	60	49 (12)	117 (43, 231)
Single lung (63)	18	3	42	52 (11)	104 (5, 194)
Double lung (10)/heart–lung	15	4	18	41 (11)	147 (94, 305)
Pulmonary hypertension (68)	25	2	41	37 (10)	173 (81, 364)
Other (21)	2	3	16	41 (12)	229 (40, 498)
Overall (653)	184	48	425	39 (14)	178 (65, 432)

Charman SC et al, *JHLT* 2002

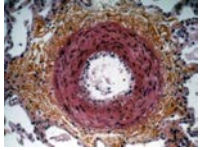
A consensus document for the selection of lung transplant candidates: 2014—An update from the Pulmonary Transplantation Council of the International Society for Heart and Lung Transplantation



Weill D et al, *JHLT* 2015

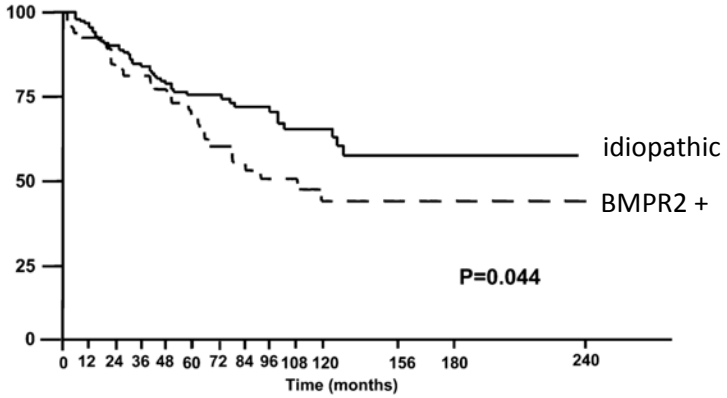
Timing for listing

- NYHA FC III or IV despite a trial of at least 3 months of combination therapy, including prostanoids
- Cardiac index of < 2 L/min/m^2
- Mean right atrial pressure of > 15 mm Hg
- 6-min walk test of < 350 m
- Development of significant hemoptysis, pericardial effusion, or signs of progressive right heart failure, including renal insufficiency, rising bilirubin, brain natriuretic peptide, or recurrent ascites.



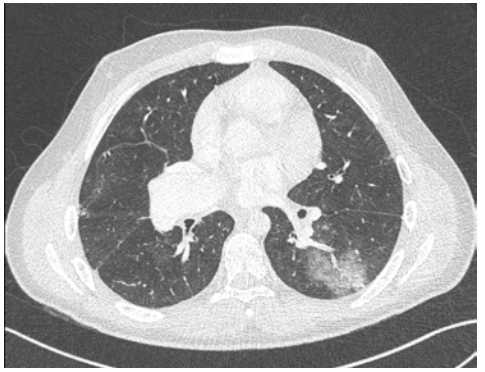
Early referral-
Etiology of pulmonary hypertension

BMPR2 vs idiopathic



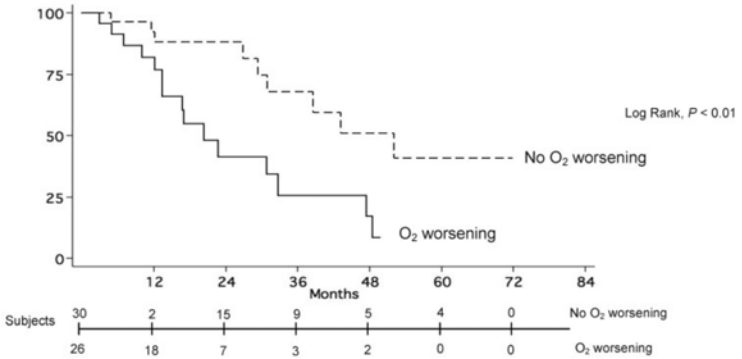
Sztrymf B et al, *AJRCCM* 2008

Hemoptysis



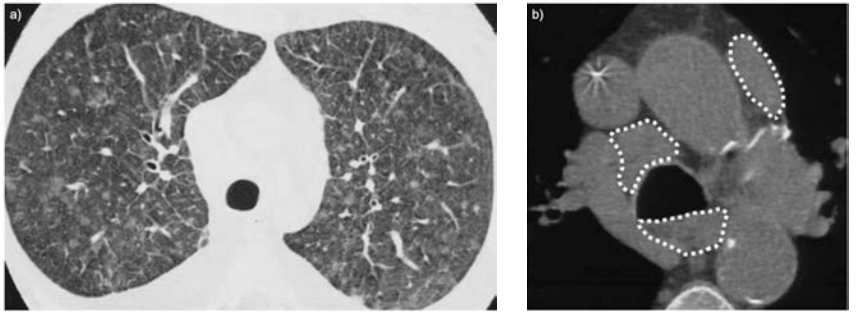
Jaïs X et al, *ATS* 2009

SSc-PAH ± ILD



Le Pavec J et al, *A&R* 2011

PVOD



Montani D et al, *ERJ* 2010

A consensus document for the selection of lung transplant candidates: 2014—An update from the Pulmonary Transplantation Council of the International Society for Heart and Lung Transplantation

Weill D et al, *JHLT* 2015

Contre-indications absolues

- Pathologies malignes < 2 ans sauf carcinomes basocellulaires. Une période de rémission ≥ 5 ans prudent. Carcinome bronchioloalvéolaire
- Dysfonction sévère d'un organe dominant (cœur, foie, rein) en l'absence de transplantation combinée
- Atteinte coronaire non accessible à une revascularisation percutanée ou associée à une dysfonction VG sévère. Transplantation cœur poumon pourrait constituer une option chez des candidats sélectionnés.
- Infection chronique incurable (HVB et HVC actives et HIV*)
- Déformations majeures de la cage thoracique
- Absence prévisible d'adhésion / compliance au projet : pathologie psychiatrique, isolement social, addiction [persistante ou sevrage < 6 mois (tabac, alcool, narcotique)]

* **1ère transplantation VIH en France à ML en 2015**

A consensus document for the selection of lung transplant candidates: 2014—An update from the Pulmonary Transplantation Council of the International Society for Heart and Lung Transplantation

Weill D et al, *JHLT* 2015

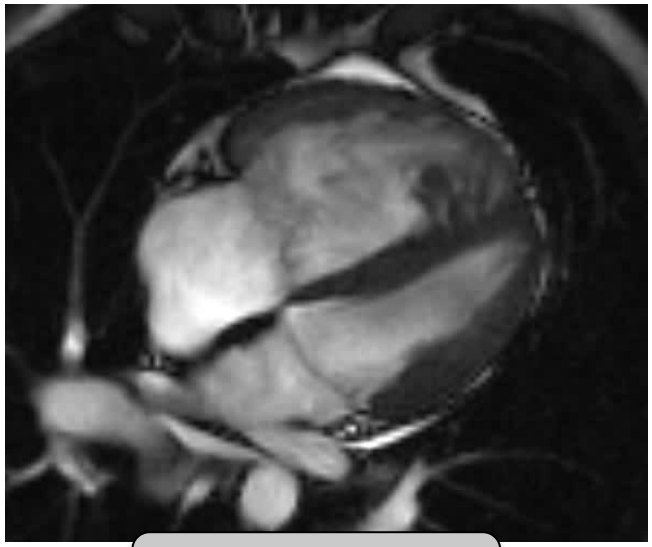
Contre-indications relatives

- Age > 65 ans
- Altération profonde de l'état général (BMI < 17 kg/m^2)
- Colonisation à bactérie, agent fungique ou mycobactérie avec haut niveau de résistance
- Obésité avec BMI > 30 kg/m^2
- Diabète, HTA, RGO, insuffisance coronaire qui doivent être médicalement optimisés avant transplantation.

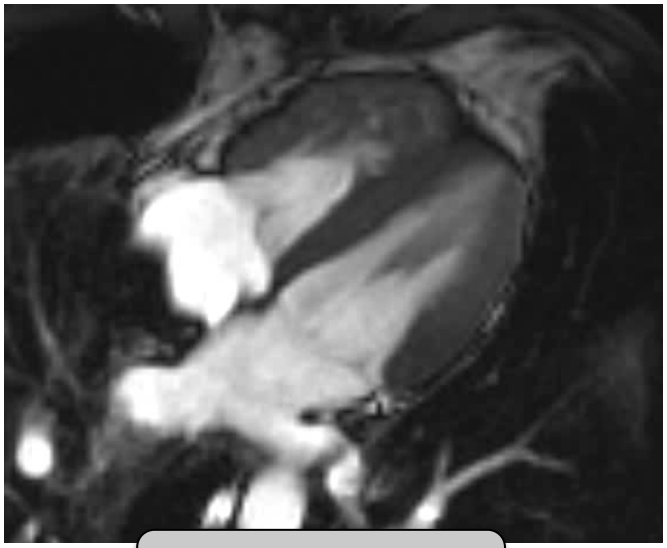
- ATCD chirurgie thoracique : augmentent difficultés techniques, risque de saignement
- Dépendance au ventilateur : pas une contre-indication si défaillance isolée et stable

Type de procédure
TBP vs TCP

Endarteriectomies des AP



Avant



Après

- La dysfonction VD récupère quelle que soit la sévérité de la dysfonction
- La récupération sera complète si la circulation artérielle pulmonaire est restaurée
- La récupération est progressive de jours à semaines

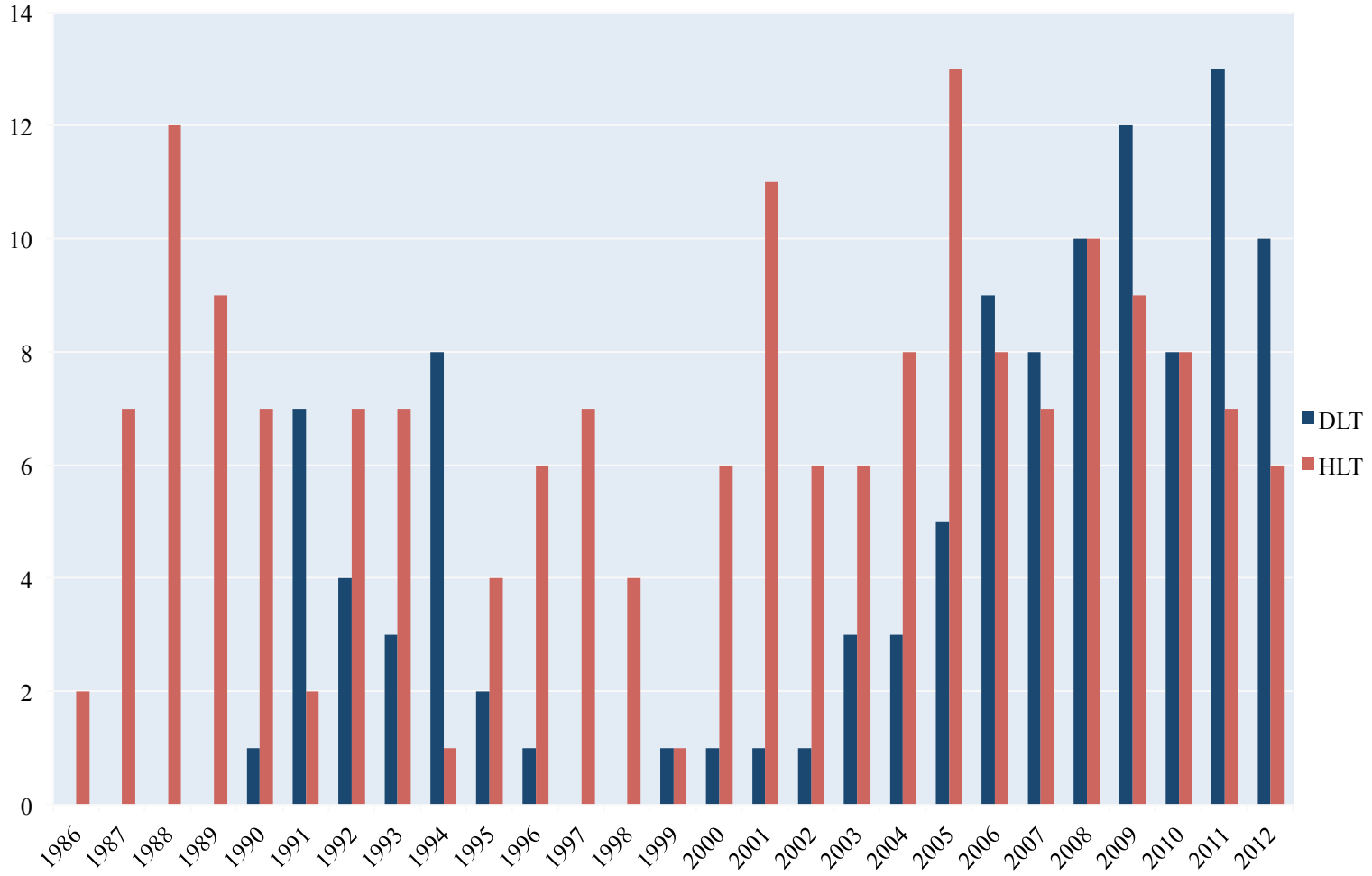
Should we perform bilateral-lung or heart–lung transplantation for patients with pulmonary hypertension?

Anne Olland[a], Pierre-Emmanuel Falcoz[a,*], Mathieu Canuet[b] and Gilbert Massard[a]

- Méta analyse portant sur 77 abstracts et 9 études
- Absence de bénéfices TCP > TBP
- TCP procédure de choix en cas de cardiopathie congénitale et de dysfonction VD et/ou VG sévère
- Pour les autres indications, TBP est associée à des résultats similaires à la TCP et offre l'avantage d'un meilleur partage des organes à transplanter
- Cut off optimal de la dysfonction VD à identifier (Fej VD 15 - 20%)

Olland A et al, Interact Cardiovasc Thorac Surg 2013

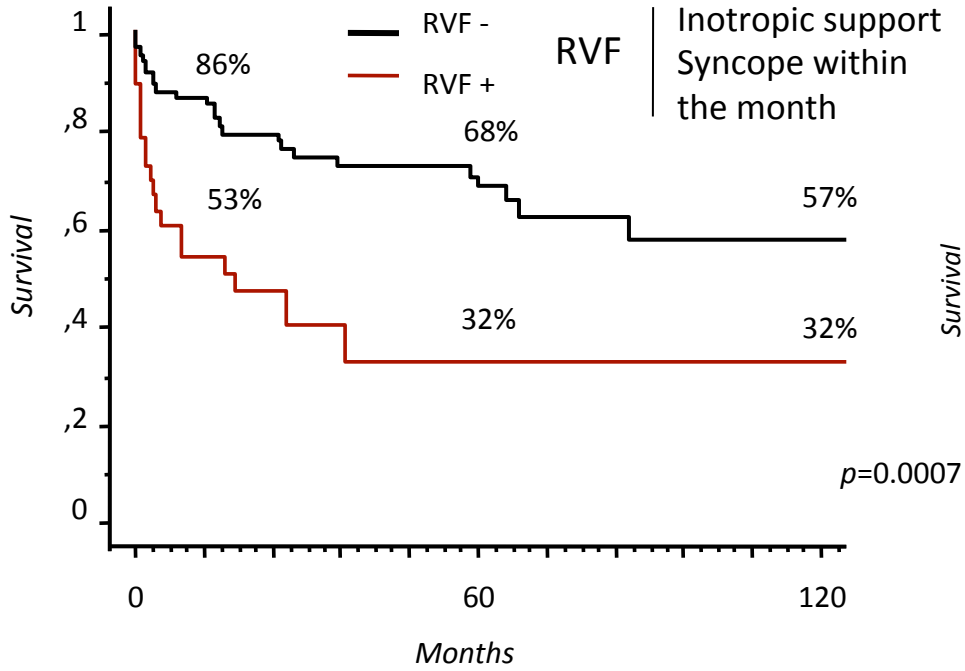
Marie Lannelongue experience with PH 1986 – 2013, *n = 295*



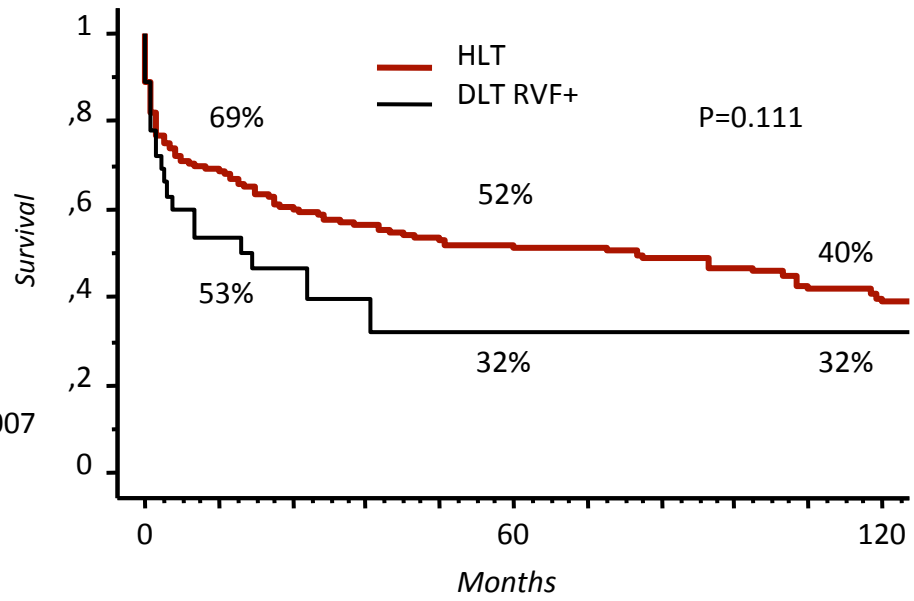
Dysfonction VD
Pronostic précoce

Marie Lannelongue experience with PH 1986 – 2013, *n = 295*

DLT ± RVF

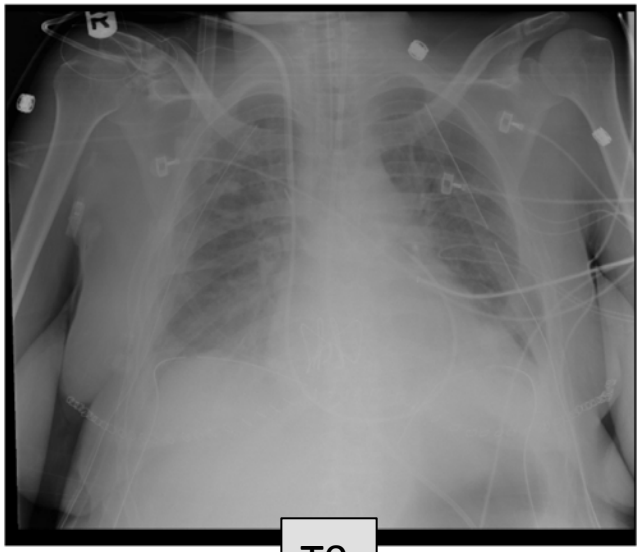


HLT vs DLT + RVF

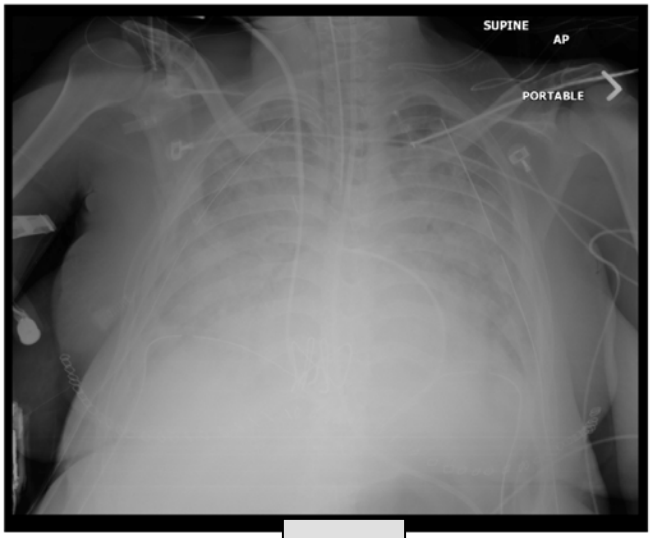


Dysfonction Primaire du Greffon

Présentation



T0



T72

Grade at T0, T24, T48, T72	Radiographic infiltrates consistent with diffuse pulmonary edema	PaO_2:FiO_2	Specific exceptions
0	–	Any	
1	+	> 300	On nasal cannula or $FiO_2 < 0.3$
2	+	200–300	
3	+	< 200	Any patients on ECMO or on NO with $FiO_2 > 0.5$ MV

Suzuki Y et al, *Semin Respir Crit Care Med* 2013

PGD risk factors

Donor inherent variables
- 21 > age > 45 yo
- African american race
- Female gender
- History of smoking

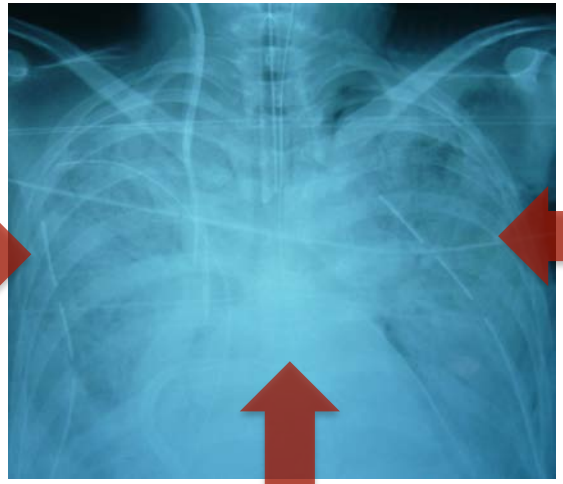
Recipient variables
- Obesity
- Female gender
- Pulmonary hypertension
- Pulmonary fibrosis
- Elevated pulmonary pressure at the time of sugery

Primary Graft Dysfunction

Donor acquired variables
- Prolonged mechanical ventilation
- Aspiration
- Head trauma
- Hemodynamic instability after brain death

Operative variables
- Single LT
- Prolonged ischemic time
- Cardiopulmonary bypass
- High FiO2

DPG dans le contexte HTAP



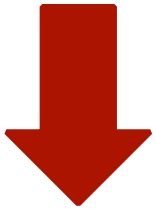
Hypertrophie VD

Septum paradoxal
Dysfonction diastolique du VG

Effondrement de la post charge VD par TBP

SUMMARY – RVD in PGD

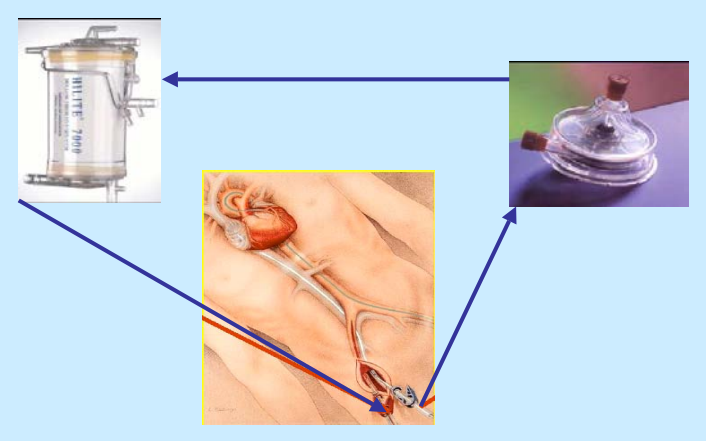
Preload optimization	Adequate perfusion	Decrease RV afterload	Ventilatory support
Low volume loading Avoid RV overload Wedge pressure CVP Echo ! Extubation ++	Norepinephrine Consider Inotropic or Inodilator support	Avoid Hypoxemia and hypercarbia Inhaled NO	Protective Strategy Low tidal volume Optimized PEEP



Failure

ECMO

Extra Corporeal Life Support



ECMO = Extracorporeal Membrane Oxygenation

Decarboxylation = ECMO low-flow

Oxygénation et decarboxylation = ECLA (ExtraCorporeal Lung Assist)

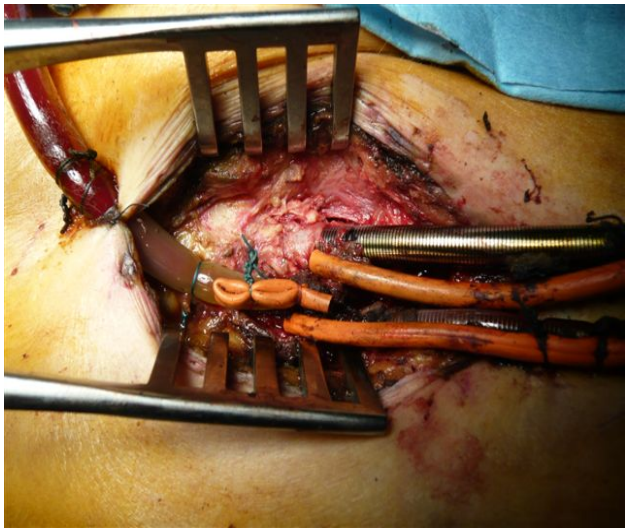
V-V, V-A, V-V-A **ECMO**

Accès Vasculaire

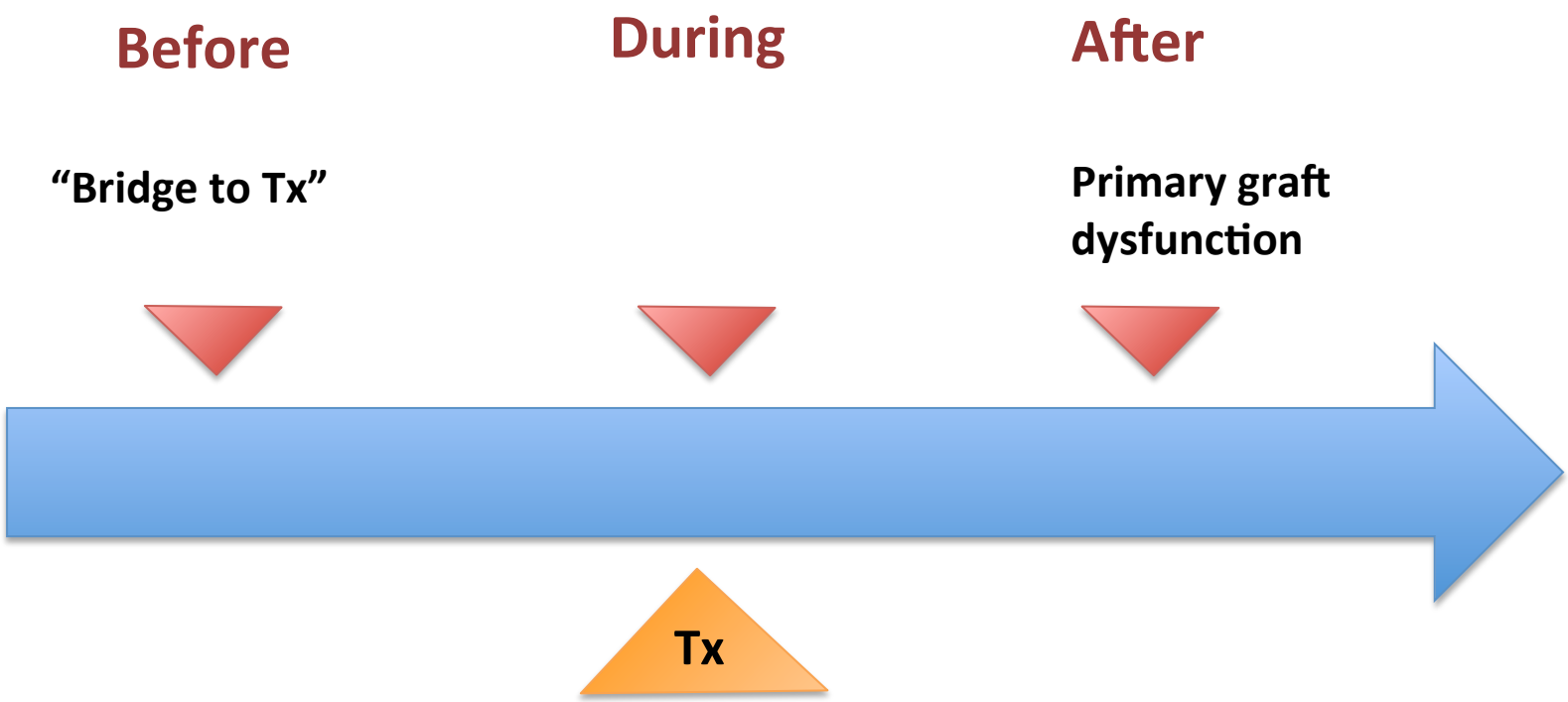
Infection

Lésions vasculaires

Hémorragie



ECMO and Lung Tx



ECMO after Tx

Preventive post-op ECMO

Keep the ECMO after Tx
Weaning process after 72h (after PGD risk)
Central or peripheral (Harlequin?)
VA-ECMO
Lung perfusion monitoring or VAV-ECMO
Possibility of awake ECMO

RHF ++
Donor / Recipient not excellent

Weaning ECMO with LA pressure monitoring

Objective criteria to wean/keep the ECMO at the end of the Tx
Allow fast post Tx recovery
Avoid prolonged ECMO complications
Allow weaning at the end of the Tx (preferential use of central ECMO)

RHF --
Donor / Recipient excellent

HTAP
Cardiopathies congénitales

REVIEW

Pulmonary arterial hypertension associated with congenital heart disease

Michele D'Alto* and Vaikom S. Mahadevan#

TABLE 1 Clinical classification of congenital, systemic-to-pulmonary shunts associated with pulmonary arterial hypertension (PAH)

Group A. Eisenmenger's syndrome

Eisenmenger's syndrome includes all systemic-to-pulmonary shunts due to large defects leading to a severe increase in PVR and resulting in a reversed (pulmonary-to-systemic) or bidirectional shunt. Cyanosis, erythrocytosis and multiple organ involvement are present.

Group B. PAH associated with systemic-to-pulmonary shunts

In these patients with moderate-to-large defects, the increase in PVR is mild-to-moderate, systemic-to-pulmonary shunt is still largely present and no cyanosis is present at rest.

Group C. PAH with small defects

In cases with small defects (usually VSD <1 cm and ASD <2 cm of effective diameter assessed by echocardiography), the clinical picture is very similar to idiopathic PAH.

Group D. PAH after corrective cardiac surgery

In these cases, CHD has been corrected but PAH is either still present immediately after surgery or has recurred several months or years after surgery in the absence of significant post-operative residual congenital lesions or defects that originate as a sequelae to previous surgery.

D'Alto M Eur Respir Review 2012

REVIEW

Pulmonary arterial hypertension associated with congenital heart disease

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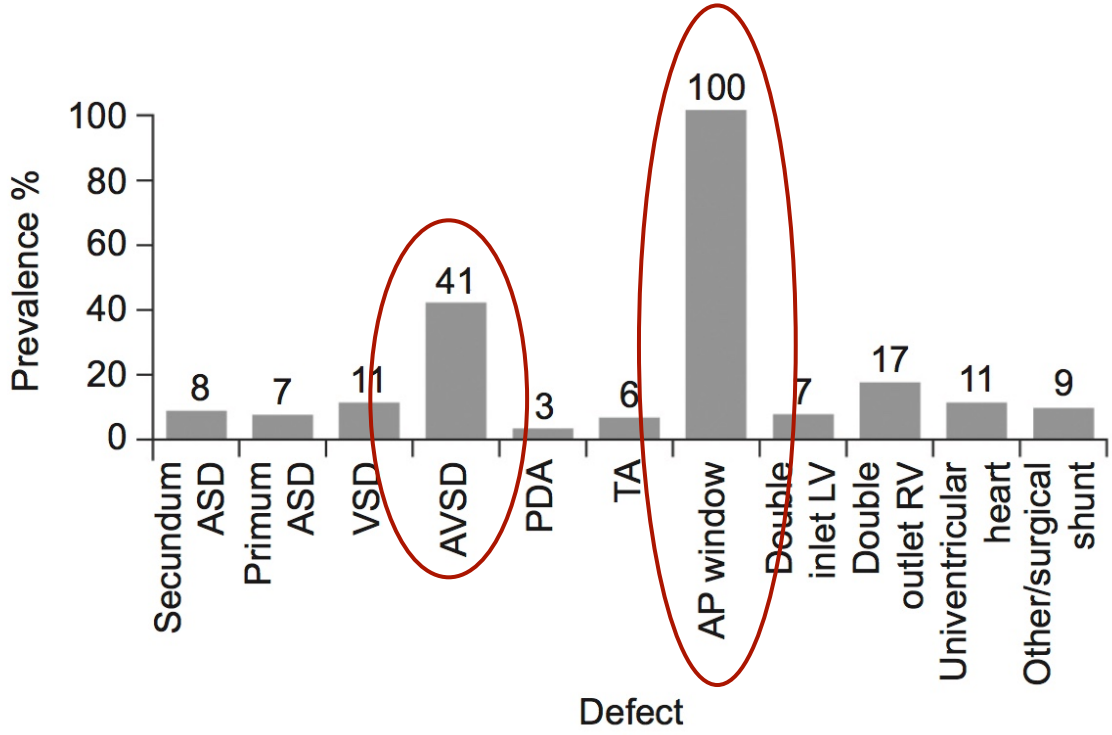


Caractérisation des lésions

- **Echo préop**
  CIA
  CIV

- **Scanner thoracique**
  Canal artériel
  Aorte
  AP
  Circulation systémique
  pleurale et médiastinale

- **Aortographie**
- **Venographie**
- **Découverte peropératoire**

D'Alto M Eur Respir Review 2012

A consensus document for the selection of lung transplant candidates: 2014—An update from the Pulmonary Transplantation Council of the International Society for Heart and Lung Transplantation

Weill D et al, *JHLT* 2015

Contre-indications absolues à la transplantation pour Eisenmenger

- ATCD de chirurgie thoracique
- Développement de la circulation systémique
- Malformations complexes associées : rachis, circulation cérébrale (Moya Moya), circulation abdominale (Syndrome d'Alagille)
- Défaillance multiviscérale

Timing pour inscription

- Difficile à déterminer : cœur fonctionnant à pression élevée (phénotype fœtal)
- Qualité de vie longtemps préservée
- Survie Eisenmenger >> HTAP idiopathique
- Absence de facteurs pronostiques clairement identifiés
- Quand la défaillance cardiaque survient, souvent trop tard (défaillance multiviscérale)

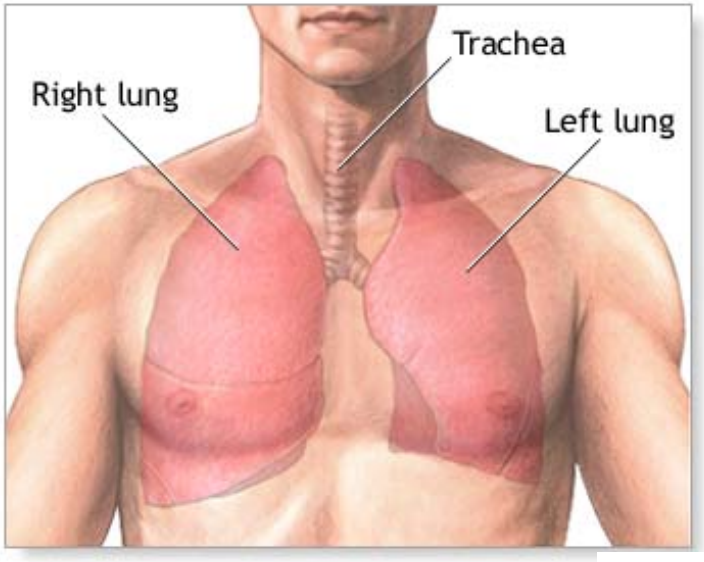
The Adult Patient with Eisenmenger Syndrome: A Medical Update after Dana Point
Part III: Specific Management and Surgical Aspects

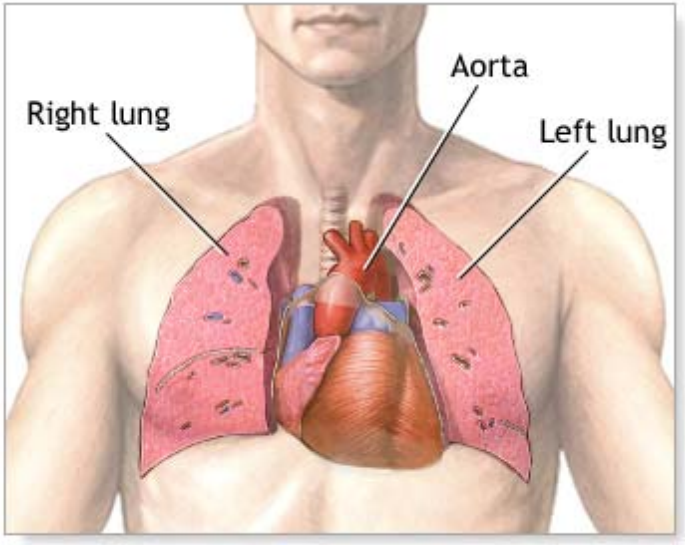
Erwin Oechslin[1,2,*], Siegrun Mebus[2], Ingram Schulze-Neick[3], Koichiro Niwa[4], Pedro T. Trindade[5], Andreas Eicken[2], Alfred Hager[2], Irene Lang[6], John Hess[2] and Harald Kaemmerer[2]

- Complexité de l'anatomie
- ATCD fréquents sternotomie / thoracotomie
- Collatéralité circulations aortique et/ou pleuropulmonaire
- Polyglobulie / hyperviscosité
- Perturbation de l'hémostase : saignement /thrombose
- Dysfonction rénale multifactorielle
- Endocardite/ embolies paradoxales /complications neurologiques
- Hyperuricémie / ostéoarthropathie hypertrophiante

Oechslin E et al, Curr Cardiol Rev 2010

Type de procédure





TBP

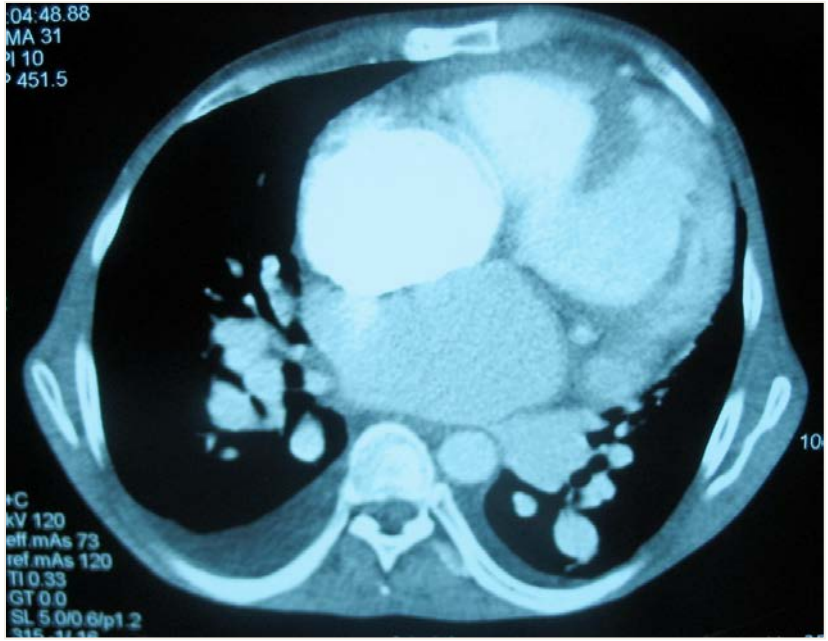
TCP

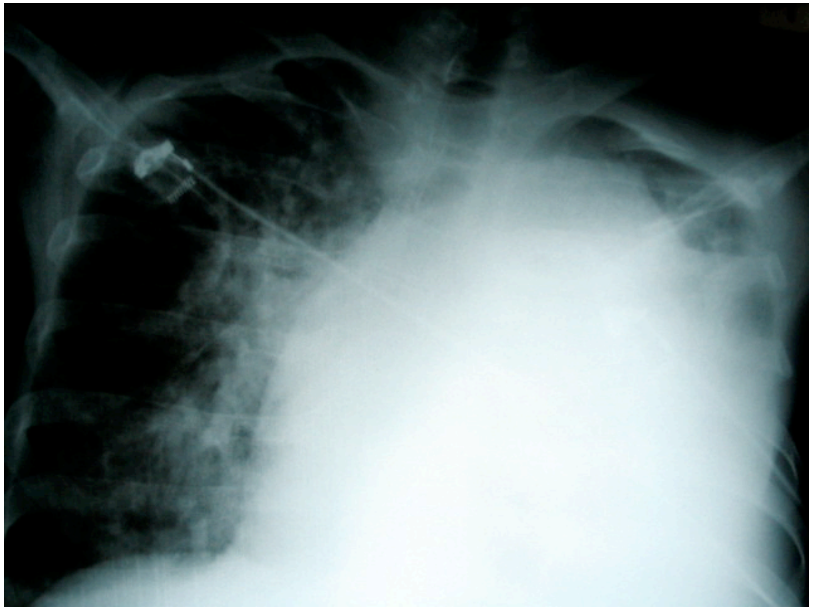
Oui

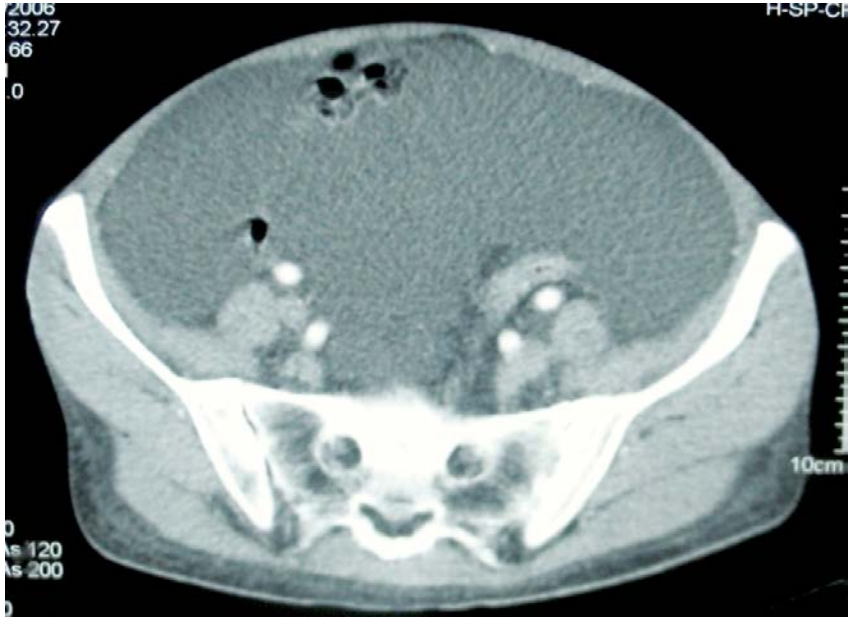
Non

- Quand malformation cardiaque corrigeable
- Quand malformation cardiaque corrigée
- Et bonne fonction VD (Fej VD ≥ 20%)

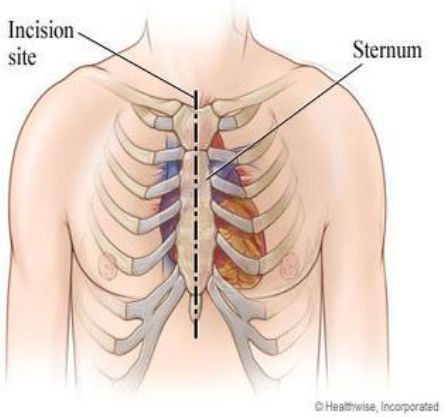
M B, 33 ans
Syndrome d'Eisenmenger
NYHA IV
O2 8 L/min
Dobutrex 10 gamma
NA 3 mg/h



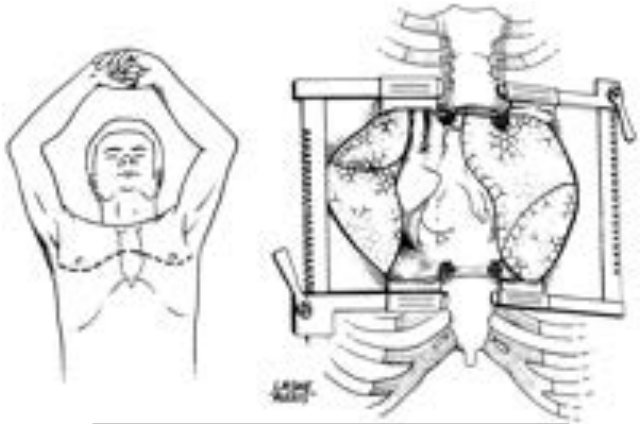




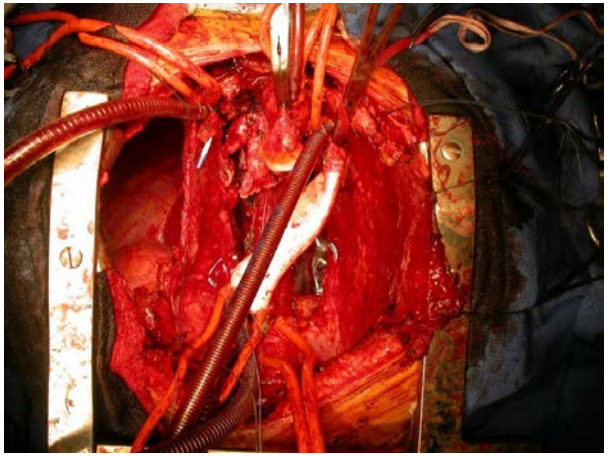
Voie d'abord

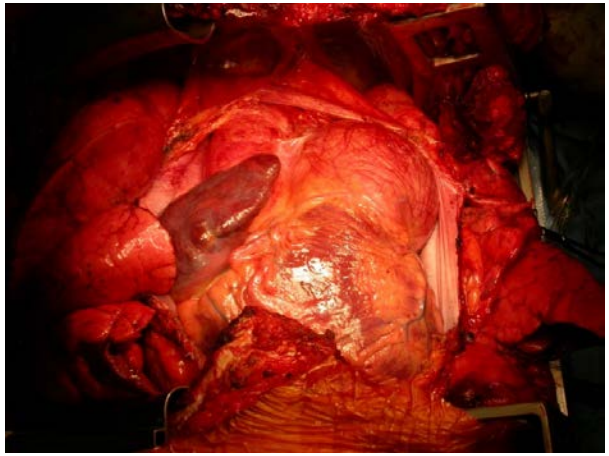


Sternotomie



Clamshell



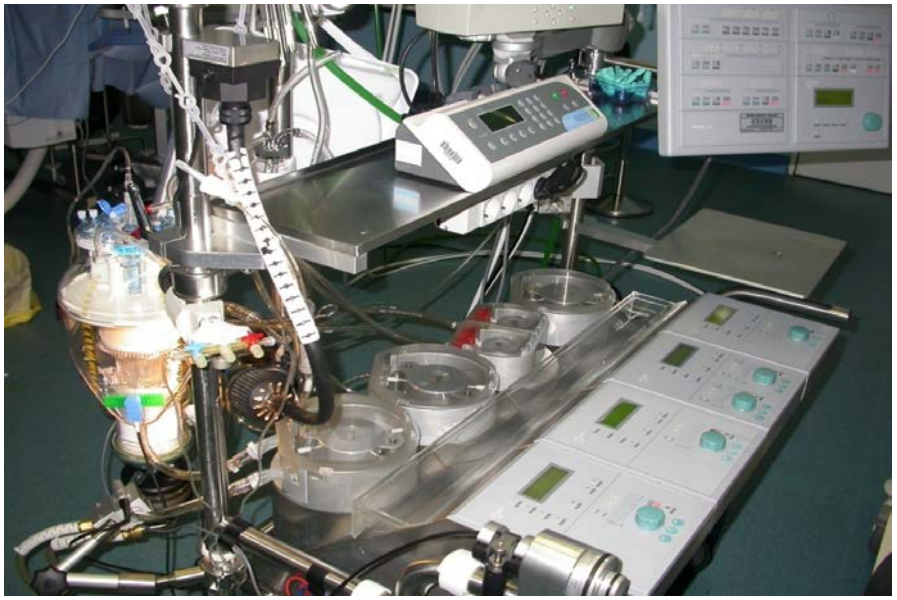


Type de procédure

	Clamshell	*Sternotomie*
Avantages	Meilleure tolérance Fermeture en 2 temps	Exposition plèvre médiastin Hémostase +++
Inconvénients	Mauvaise exposition plèvre médiastin Hémostase pleurale	Moins bonne tolérance Possible pseudarthrose

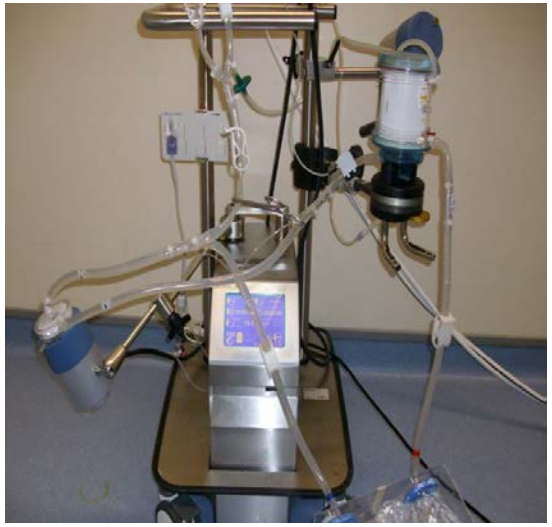
Type d'assistance

CEC



Presque toujours
Récupération saignement

ECMO



Uniquement pour TBP

Particularités de la greffe

Polyglobulie
Microglobulie

↓

Hémolyse sous CEC

↓

Insuffisance rénale
Saignement

↓

Exsanguino transfusion

Réparation chirurgicale des lésions associées

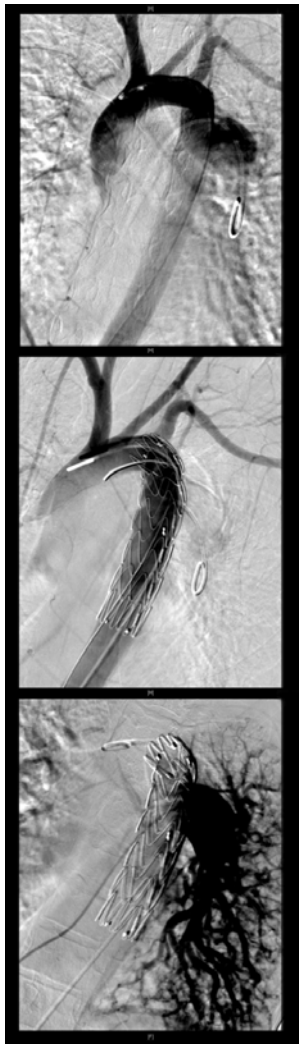
↓

CIA – Art systémique Aorte

Radiologie interventionnelle

↓

CIA – Art bronchique – Potts



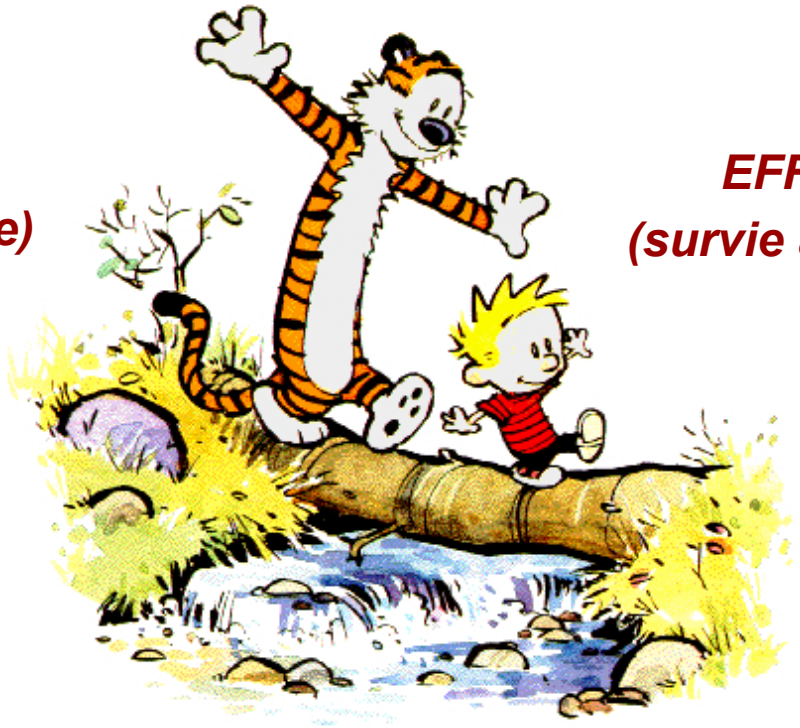
Eisenmenger synthèse

- Mortalité opératoire élevé 20-30%
- Réservée à des centres ultraspécialisés
- Approche pluridisciplinaire cardiopédiatres / pneumologues / chirurgiens
- Survie à long terme après cap aigu de 50 à 70% à 5 ans et 30 à 50% à 10 ans

Programme
de super U pour HTAP

Répartition *des organes*

EQUITE
(décès sur liste attente)



EFFCIENCE
(survie après greffe)

PROCEDURES d'application des REGLES de REPARTITION et d'ATTRIBUTION des GREFFONS PRELEVES sur PERSONNE DECEDEE

Application de l'Arrêté du 06 novembre 1996 modifié par l'arrêté du 30 août 2002 et du 18 Juin 2004.

« Les règles de répartition et d'attribution de ces greffons doivent respecter les principes d'équité, l'éthique médicale et viser l'amélioration de la qualité des soins…..Ces règles font référence aux notions de priorité et de dimension territoriale. Ces notions traduisent le souci de rechercher l'équilibre entre une répartition la plus équitable possible et les contraintes techniques inhérentes au prélèvement, au transport et au maintien de la qualité des greffons.

L'objectif de ces règles est de tenir compte de l'urgence de la greffe ou de la difficulté particulière d'y accéder pour certains malades, tout en recherchant l'utilisation optimale des greffons. L'évaluation des conséquences de ces règles sur la durée d'attente des malades et les résultats des greffes permettra leur amélioration au fur et à mesure des progrès techniques.

Répartition *des organes*

Répartition nationale

SU CP
±dérogation de groupe
8 j prolongée 1 fois

2006

pas de limite de temps
pour receveurs pédiatriques

SU-P
±dérogation de groupe
Critères d'inclusion spécifiques
selon catégories
Muco-DDB
Fibrose pulmonaire
Maladie vasculaire pulmonaire
8 jours prolongation 1 fois

2007

pas de limite de temps
pour receveurs
pédiatriques

Priorité nationale pédiatrique
donneurs <55 ans et <50 kg

2005

Greffes simultanées
Poumon + autre organe

Attribution géographique
locale
régionale
nationale

Urgences régionales

Règle d'attribution *prioritaire*

1. MUCOVISCIDOSE ET DDB (DILATATION DES BRONCHES)

PATIENT SOUS VENTILATION INVASIVE (INTUBATION) AVEC/SANS ASSISTANCE TYPE ECMO[1]

OU MENACE DE VENTILATION INVASIVE : VNI[2] > 18HEURES/J DEPUIS ≥ 3 JOURS ET PACO2 > 80 MMHG SOUS VNI EN L'ABSENCE DE CAUSE REVERSIBLE

OU MISE SOUS ASSISTANCE TYPE ECMO

2. FIBROSE PULMONAIRE IDIOPATHIQUE OU SECONDAIRE

PATIENT SOUS VENTILATION INVASIVE (INTUBATION) AVEC/SANS ASSISTANCE TYPE ECMO

OU MENACE DE VENTILATION INVASIVE : OXYGENOTHERAPIE > 12L/MN ET SAO2 AU MASQUE < 90% MALGRE TRAITEMENT MEDICAL MAXIMAL (BOLUS SOLUMEDROL, …) EN L'ABSENCE DE CAUSE REVERSIBLE

OU MISE SOUS ASSISTANCE TYPE ECMO

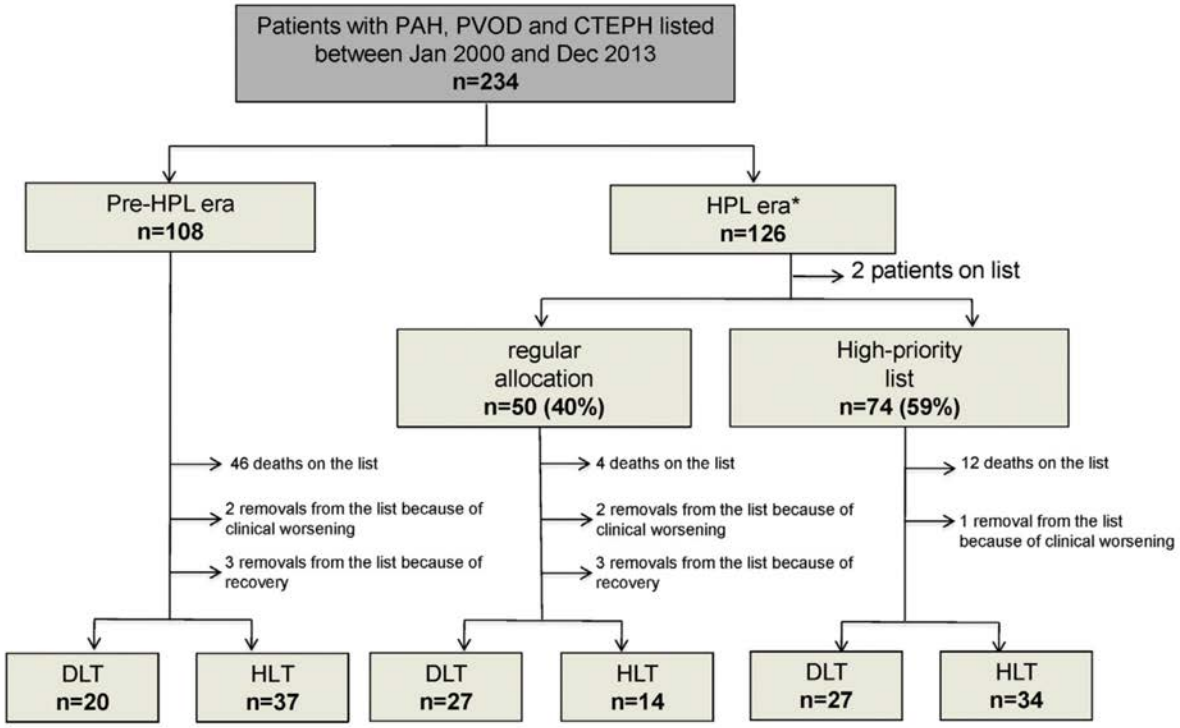
3. MALADIES VASCULAIRES PULMONAIRES

PATIENT PRESENTANT UNE HYPERTENSION PULMONAIRE SEVERE NE S'AMELIORANT PAS APRES PLUS DE 72 HEURES D'UN TRAITEMENT MEDICAL MAXIMAL INCLUANT L'ADMINISTRATION CONTINUE D'INOTROPES EN UNITE DE SOINS INTENSIFS ET/OU DE PLUSIEURS DES TRAITEMENTS SPECIFIQUES DE L'HYPERTENSION PULMONAIRE.

L'HYPERTENSION PULMONAIRE SEVERE EST DEFINIE PAR L'ASSOCIATION D'UN STADE IV DANS LA CLASSIFICATION NYHA, D'UN INDEX CARDIAQUE INFERIEUR A 2 L/MIN/M^2 ET DES RESISTANCES ARTERIELLES PULMONAIRES SUPERIEURES A 1200 DYN.SEC.CM^{-5}

Impact of High-Priority Allocation on Lung and Heart-Lung Transplantation for Pulmonary Hypertension

Laurent Savale, MD, PhD, Jérôme Le Pavec, MD, PhD, Olaf Mercier, MD, PhD, Sacha Mussot, MD, Xavier Jaïs, MD, Dominique Fabre, MD, Caroline O'Connell, MD, David Montani, MD, PhD, François Stephan, MD, Olivier Sitbon, MD, PhD, Gérald Simonneau, MD, Philippe Dartevelle, MD, Marc Humbert, MD, PhD, and Elie Fadel, MD, PhD



* Since September 2006 for HLT and July 2007 for DLT

Savale L et al, *Ann Thorac Surg* 2017

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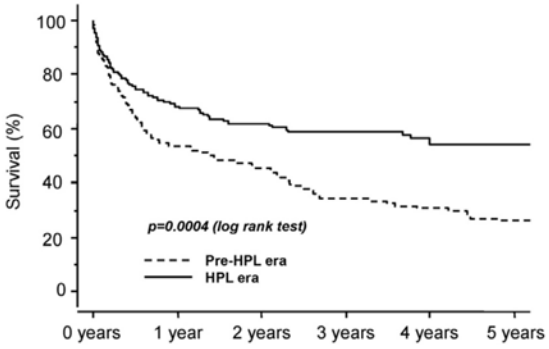
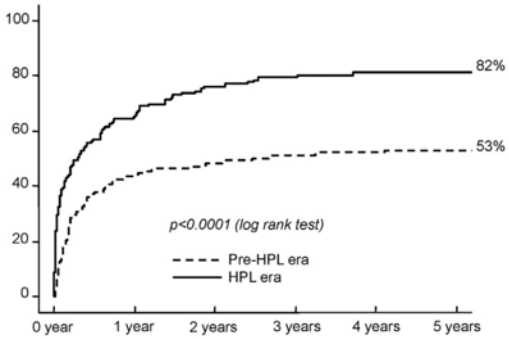
Survival after listing



Transplantation incidence



Survival after transplantation

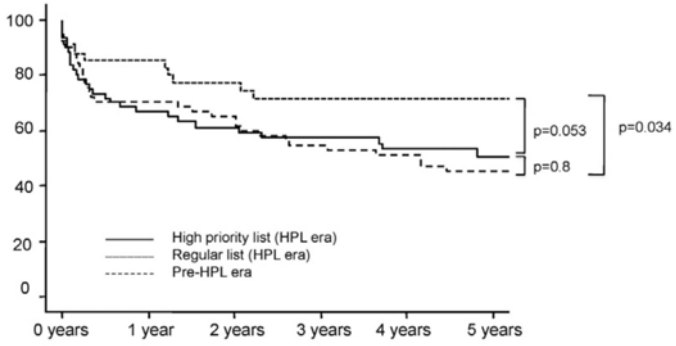


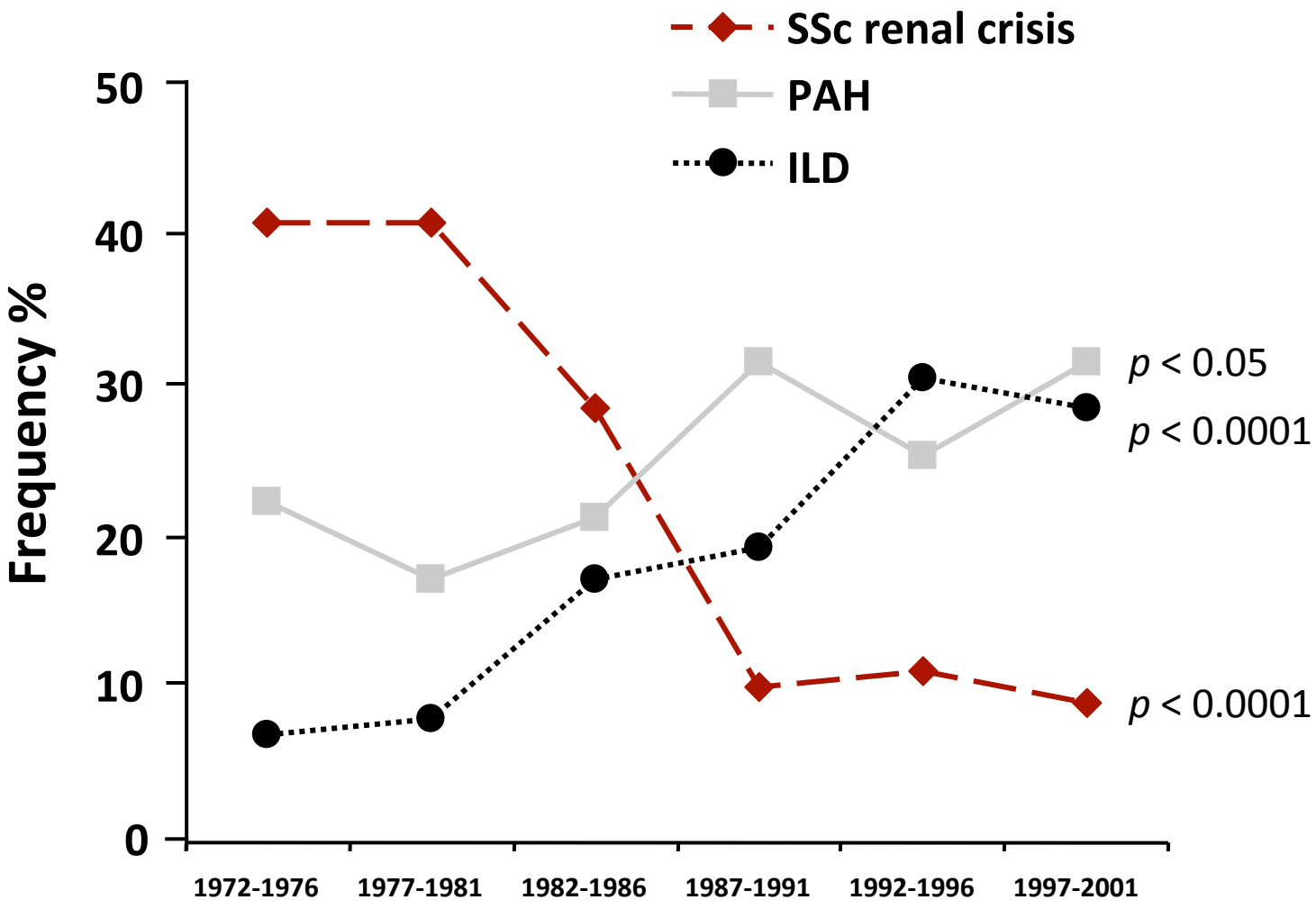
Table 3. Posttransplantation Morbidity After Implementation of the High-Priority List

Variable[a]	Regular Allocation (n = 41)	High-Priority Allocation (n = 61)	*p* Value
Follow-up duration before listing, mon	39 (16–72)	30 (5–60)	0.6
Intensive care unit length of stay, d	21 (14–28)	20 (13–38)	0.27
MV duration, d	11 (4–20)	11 (5–27)	0.94
Postoperative extracorporeal life support	11 (27)	20 (33)	0.6
Primary graft dysfunction	10 (29)	22 (36)	0.28
Patients with CLAD, %			
At 1 year	9.2	13.6	0.058
At 2 years	15.8	27	
At 3 years	19.2	32.8	
At 5 years	24.2	54.7	

Savale L et al, *Ann Thorac Surg* 2017

Transplantation pour sclérodermie

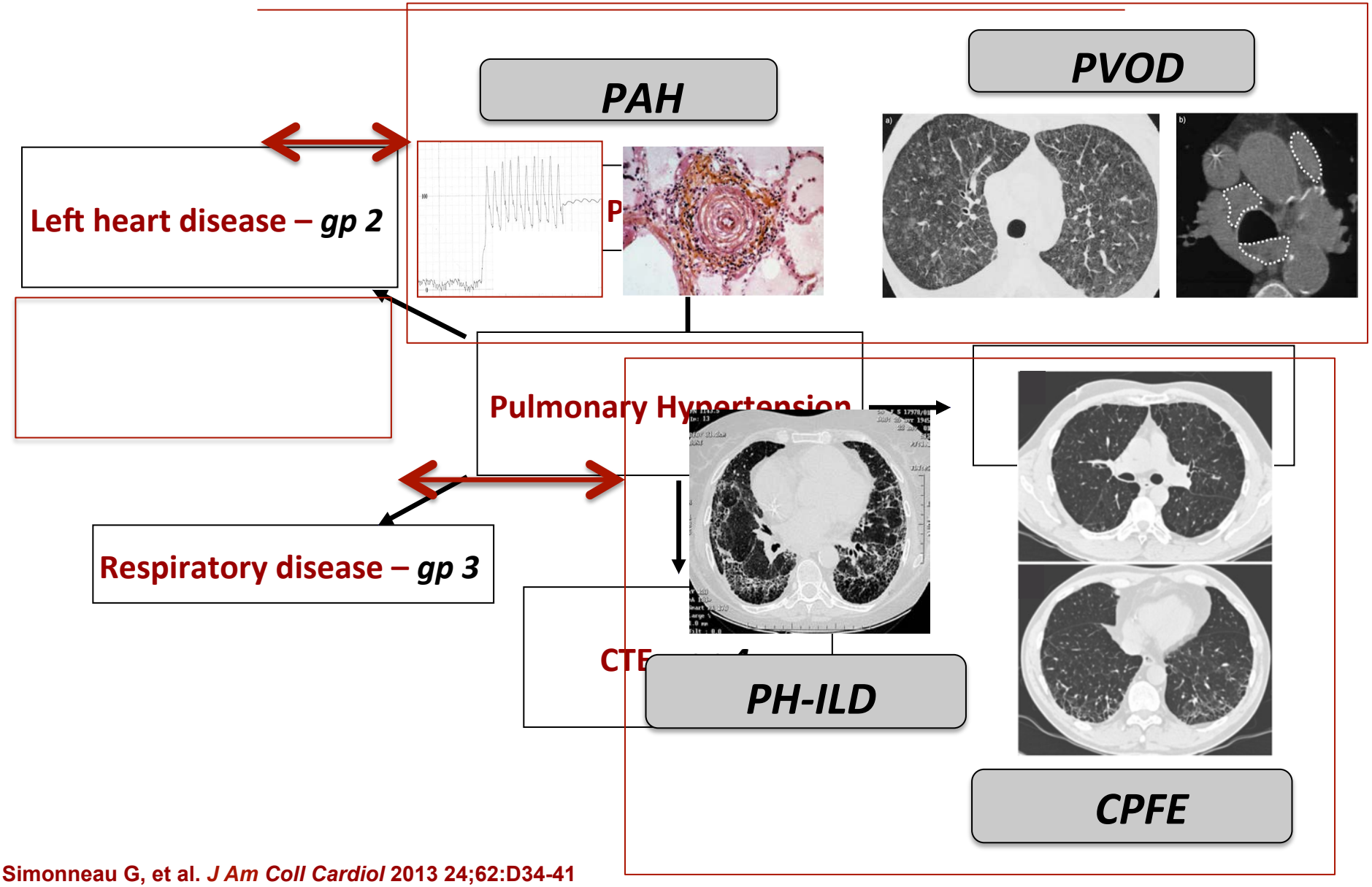
Pulmonary involvement is the major cause of death in SSc



Steen VD and Medsger TA. *Ann Rheum Dis* 2007; 66:940-4.

Updated classification of Pulmonary Hypertension

Spectrum of pulmonary hypertension in SSc



Simonneau G, et al. *J Am Coll Cardiol* 2013 24;62:D34-41

Lung and heart-lung transplantation for systemic sclerosis patients. A monocentric experience of 13 patients, review of the literature and position paper of a multidisciplinary Working Group

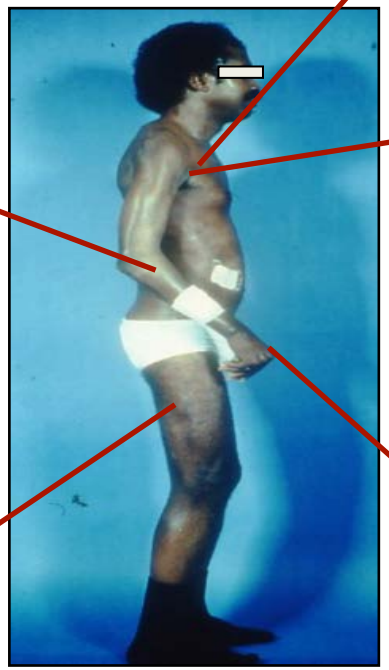
Proposed specific SSc contraindications

Left Heart

- heart:
  - conduction abnormalities and/or rhythm disturbances (symptomatic bradycardia, ventricular and atrial tachycardia): these must be managed prior to LT (implantation of a pacemaker, where appropriate) but are not a contraindication if HLT is considered.

Kidneys

- kidney:
  - renal function should have been stable for 3 months except in the case of acute functional renal failure related to right ventricle dysfunction;
  - interval < 3 years between SRC and HLT/LT;
  - increased risk of scleroderma renal crisis:
    a. diffuse systemic sclerosis evolving for less than 3 years since the first non-Raynaud sign/symptom;
    b. rapidly progressive and severe cutaneous involvement: progression of the cutaneous involvement characterised by an increase of more than 25% in Rodnan score within 6 to 12 months;
    c. corticosteroids > 15 mg prednisone (or equivalent)/day.



Oesophageal

- gastrointestinal:
  - oesophageal stricture;
  - active and severe upper gastrointestinal ulcerations despite optimal treatment, including proton pump inhibitors and prokinetics;
  - high grade dysplasia in a Barett's esophagus;
  - gastroparesis (abnormal gastric emptying [< 25% clearance at 90 min post-ingestion]) despite medical treatment;
  - chronic gastrointestinal bleeding with or without anaemia;

Fingers

- Digital ulcers:
  - > 1 severe episode/year despite optimal treatment;
  - active digital ulcer: temporary contraindication.

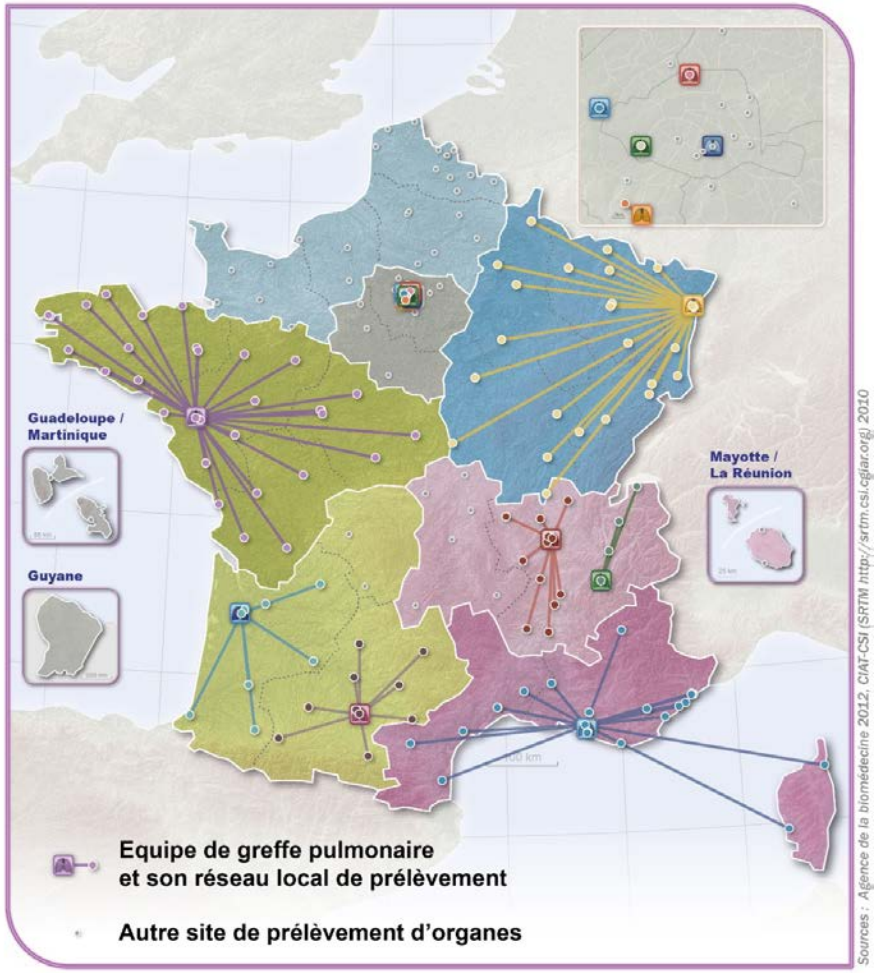
Muskuloskeletal

- Uncontrolled active inflammatory myopathy; progressive myopathy; myopathy with diaphragm involvement;

Launay D, *et al.* Press Med 2014; 43:e345-63

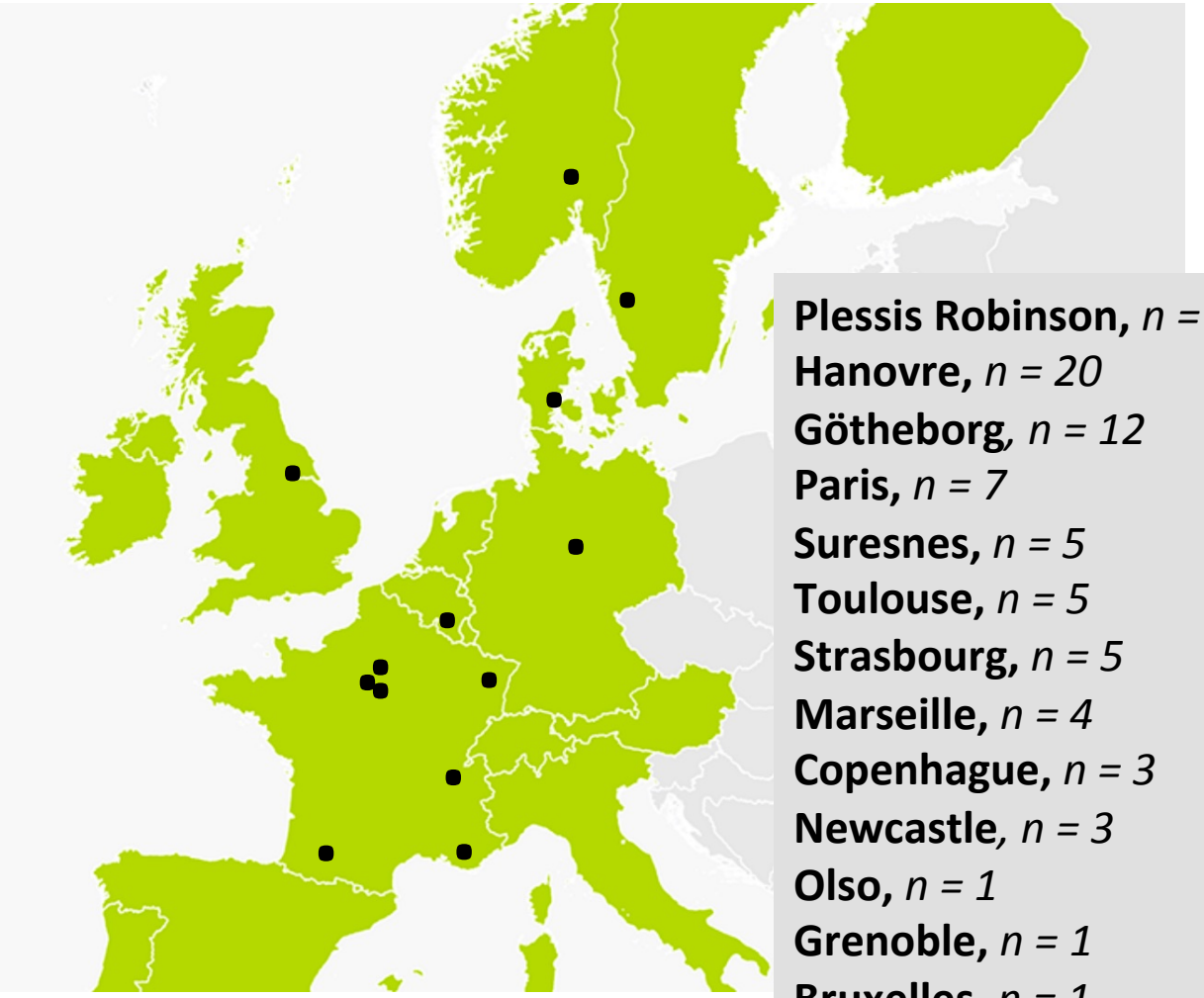
French experience of transplantation for SSc

French network of lung transplantation



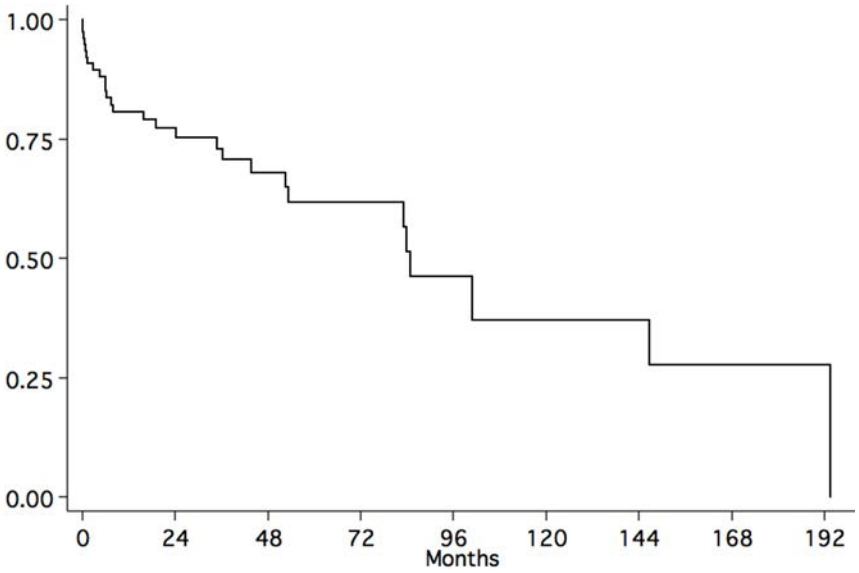
- All SSc transplanted in France
- Multicenter retrospective
- Clinical phenotypes ILD / PAH / PH-ILD / PVOD / CPFE
- Comorbidities incl GER, Left Heart & renal diseases
- Prognostic factors / Survival
- Post transplant evolution : pulmonary & renal functions, digital injuries,

European experience of transplantation for SSc, n = 92



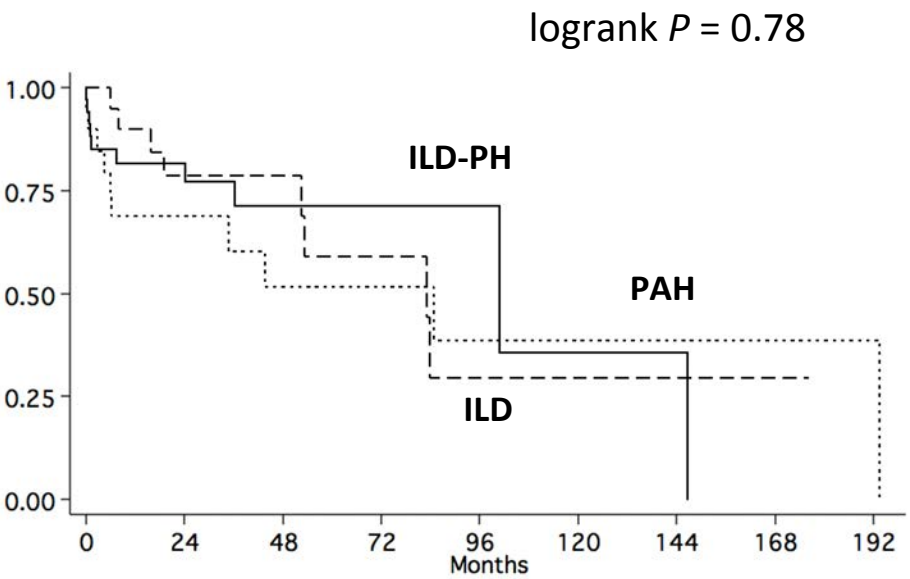
Plessis Robinson, *n = 24*
Hanovre, *n = 20*
Götheborg, *n = 12*
Paris, *n = 7*
Suresnes, *n = 5*
Toulouse, *n = 5*
Strasbourg, *n = 5*
Marseille, *n = 4*
Copenhague, *n = 3*
Newcastle, *n = 3*
Olso, *n = 1*
Grenoble, *n = 1*
Bruxelles, *n = 1*

Kaplan–Meier survival estimates from the date of transplantation of the overall population (n = 78)



Survival rates were 81%, 71%, and 62% at 1, 3, and 5 years, respectively.

Kaplan–Meier survival estimates from the date of transplantation according to SSc clinical phenotypes



Survival rates were 69, 60, and 52% in PAH, 90%, 79%, and 59% in ILD and 82, 71 and 71 % in ILD-PH patients at 1, 3 and 5 years respectively

Transplantation pour HTAP

- Transplantation pulmonaire difficile
- TBP technique de choix – TCP réservée aux cardiopathies congénitales et dysfonction VD sévères
- Pronostic précoce en rapport avec dysfonction VD / DPG
- Bénéfices de la SuperU
- Transplantation SSC possible – sélection – évaluation digestive

DU transplantation pulmonaire et cardiopulmonaire

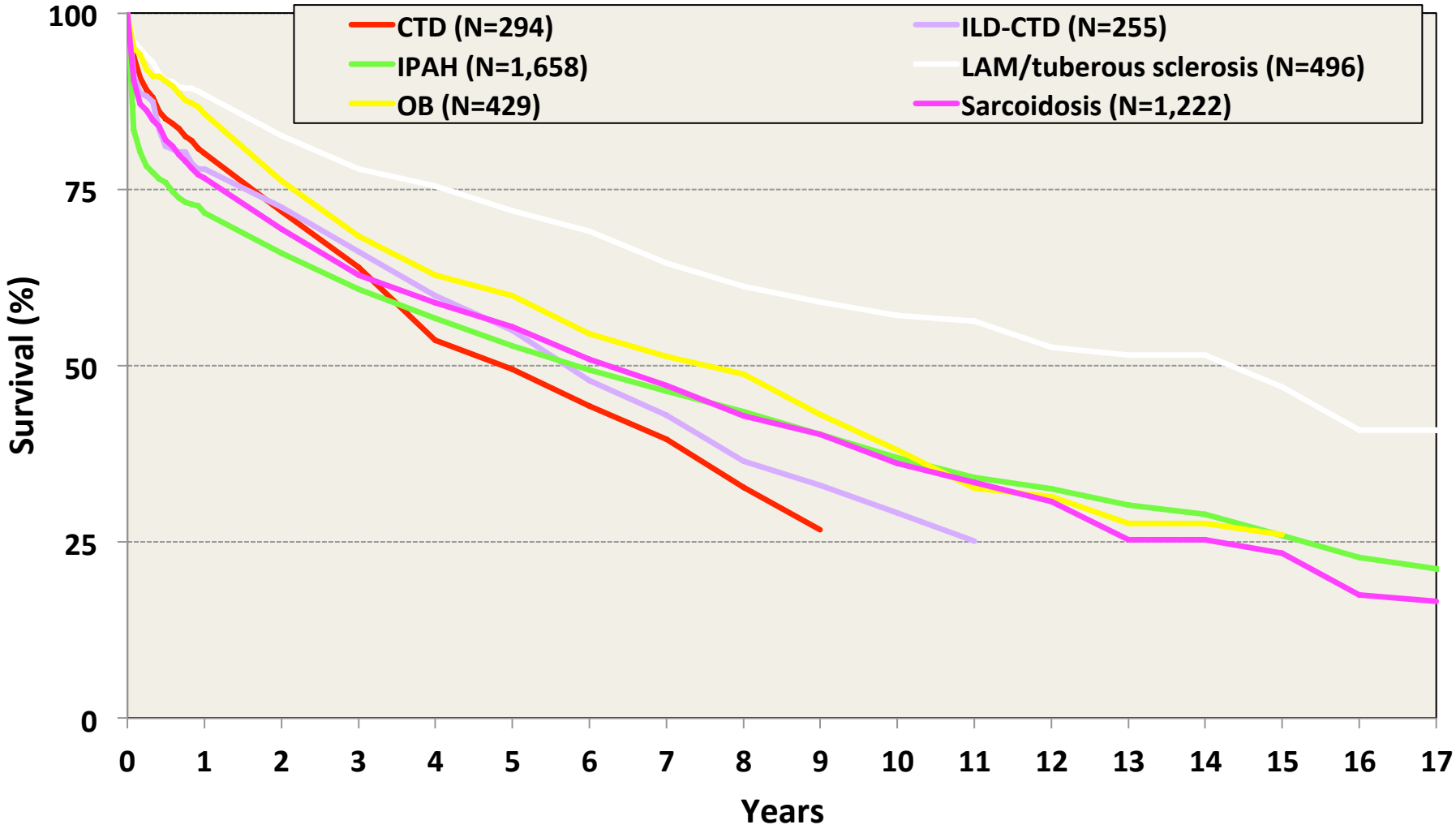
Drs Olaf Mercier & Jérôme Le Pavec

Hôpital Marie Lannelongue

m.hadjer@ccml.fr

lepavec@gmail.com

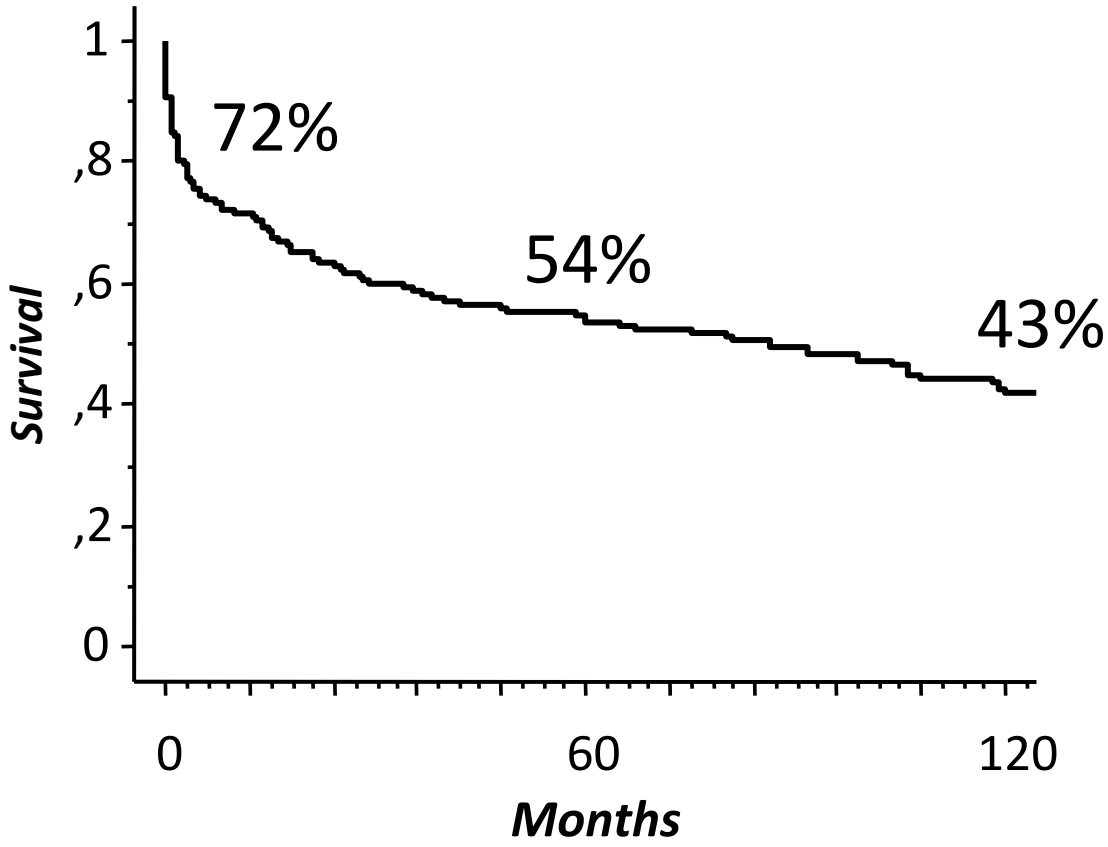
Adult Lung Transplants
Kaplan-Meier Survival by Diagnosis
(Transplants: January 1990 – June 2014)



ISHLT • INTERNATIONAL SOCIETY FOR HEART AND LUNG TRANSPLANTATION

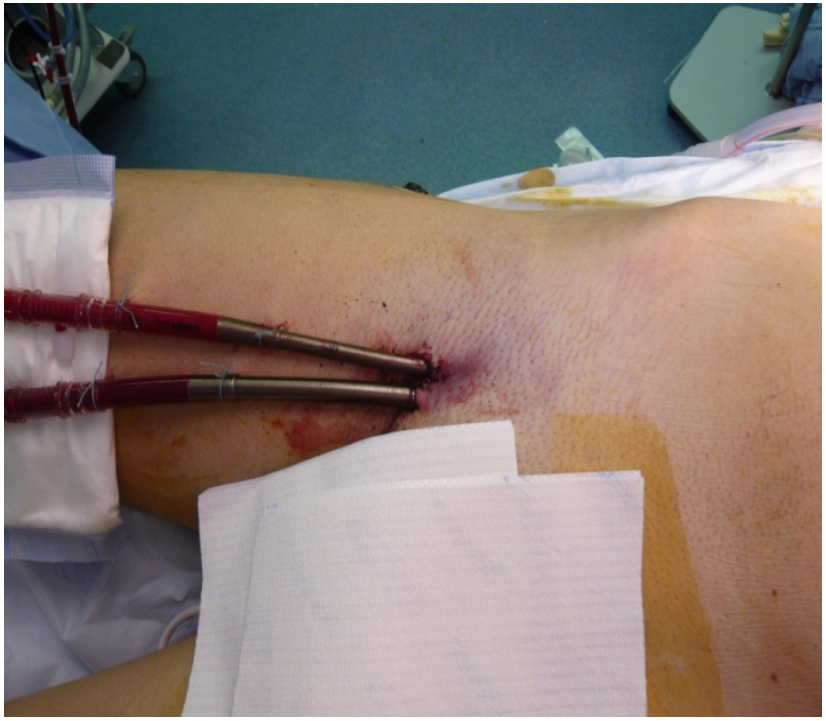
JHLT. 2016 Oct; 35(10): 1149-1205

Marie Lannelongue experience with PH 1986 – 2013, *n = 295*



ECMO before Tx

VA ECMO



Fast **percutaneous** or short open cannulation (local anest.)

Femoral or axillary artery (ambulation)
Groin **infection**

Risk of **lower limb ischemia**

ECMO during Tx

ECMO

Miniaturized, low priming
No Air-liquid contact
Less anticoagulation
Less Hemodilution
Less inflammatory response
Long-term use

No possibility for heart surgery
Active drainage
Partial heart support, no vent
Risk of air block

CBP

Higher priming volume
Air-liquid contact
Full anticoagulation
Hemodilution
Inflammatory response
Short term use (<6h)

Open heart surgery

Passive drainage
Total heart venting

Less risk of air block

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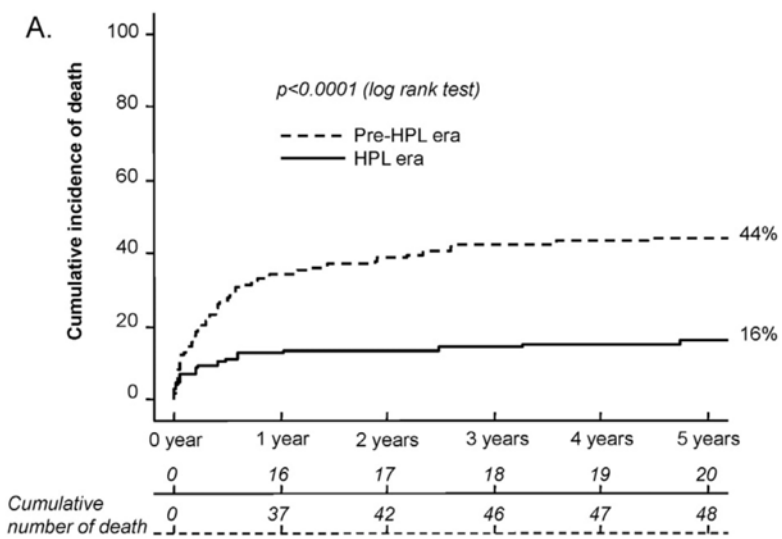

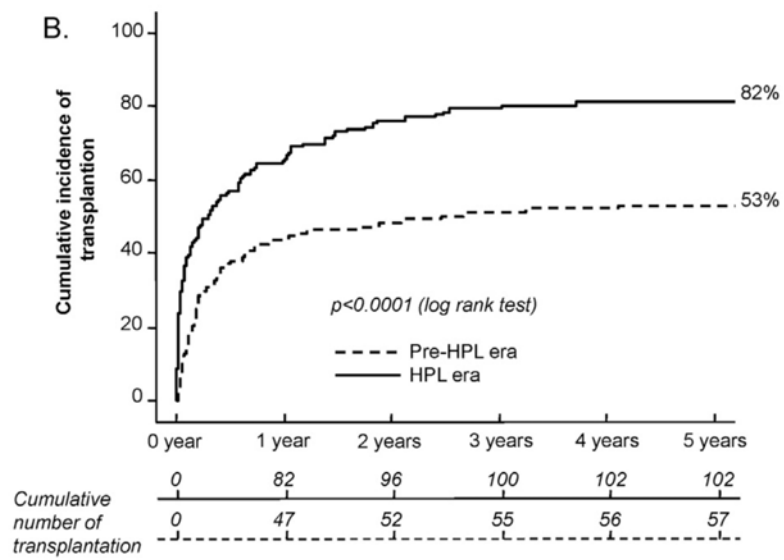

Savale L et al, *Ann Thorac Surg* 2017