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GUVERNUL ROMÂNIEI



Fondul Social European
POSDRU 2007-2013



Instrumente Structurale
2007-2013



GUVERNUL ROMÂNIEI
MINISTERUL MUNCII, FAMILIEI,
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ȘI PERSOANELOR VÂRSTNICE
OIROSDRU REGIUNEA CENTRU



UNIVERSITATEA DE MEDICINĂ ȘI
FARMACIE "CAROL DAVILA"
BUCUREȘTI

AD-COR Program inovativ de formare in domeniul cardiologiei pediatrice POSDRU/179/3.2/S/152012

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MODUL TEORETIC

Fontan operation in adulthood

Imputernicit: Prof. Dr. Tammam Youssef

Activitate prestata de I.R.C.C.S. POLICLINICO SAN DONATO – MILANO, ITALIA
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Fontan operation in adulthood

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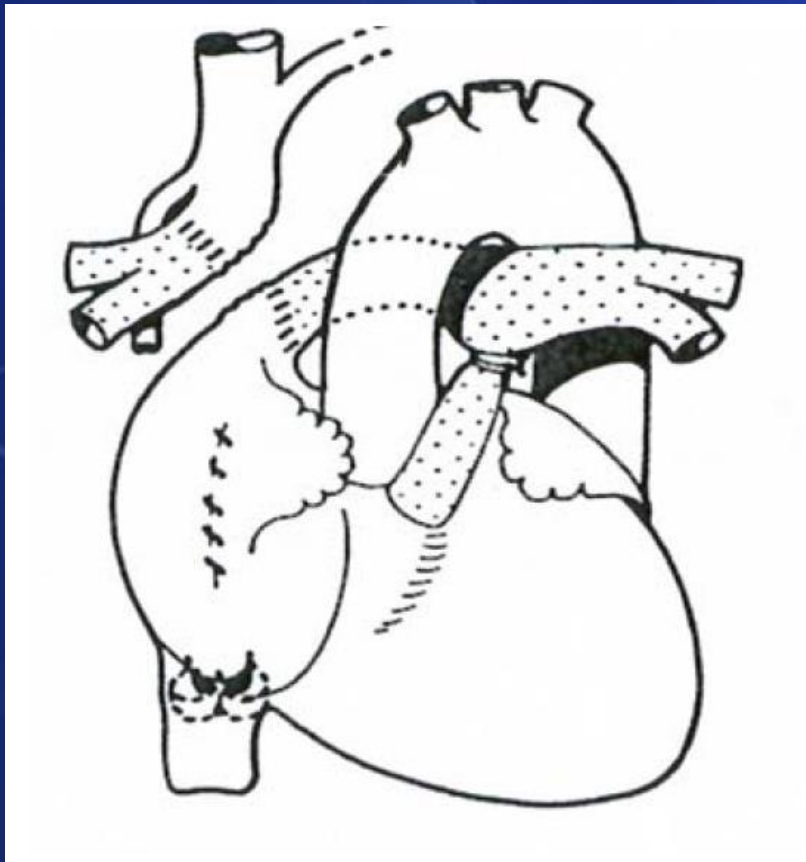
FONTAN HISTORY

- 1971 Fontan
- 1973 Kreuzer
- 1979 Bjork RA→RV Anast.
- 1980s Lateral Tunnel
- 1990s Extracardiac conduit

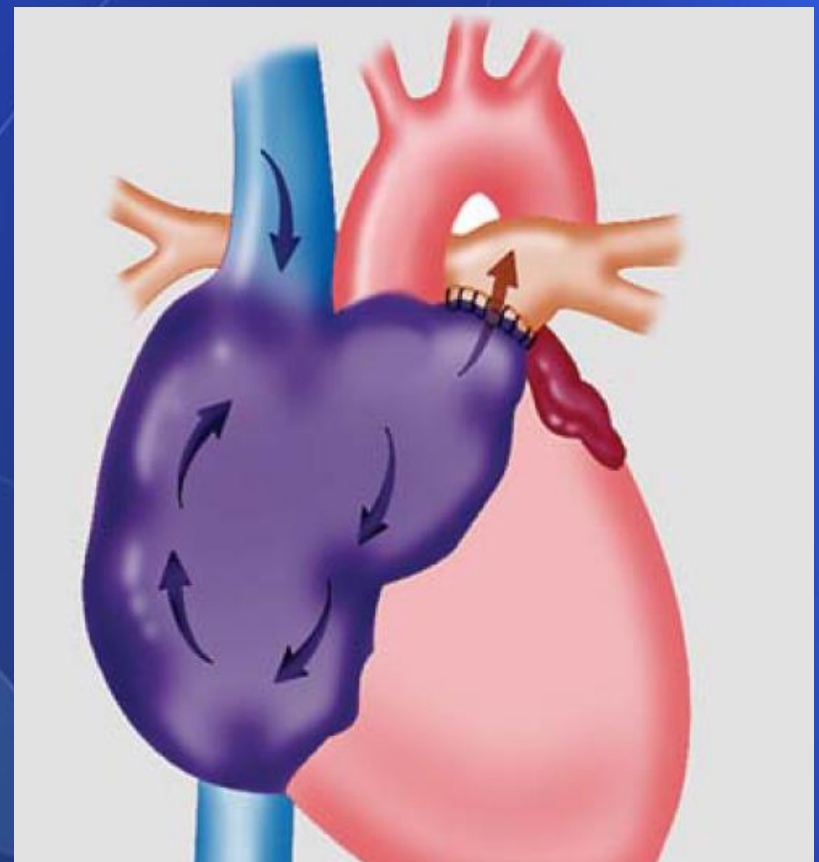
FONTAN

Surgical Techniques

The original Fontan procedure



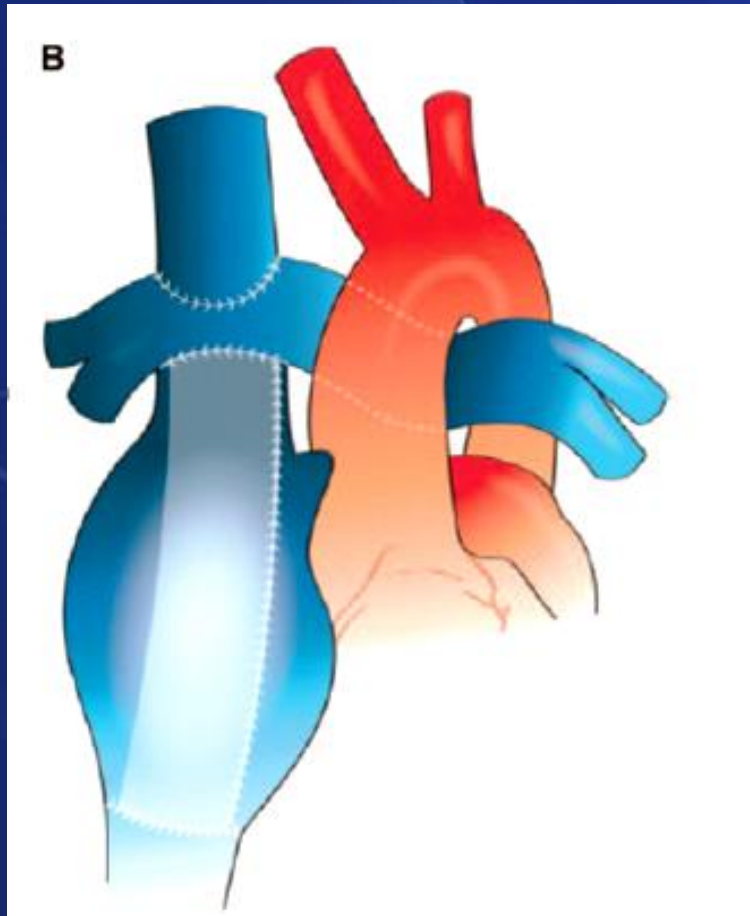
Atriopulmonary Fontan connection



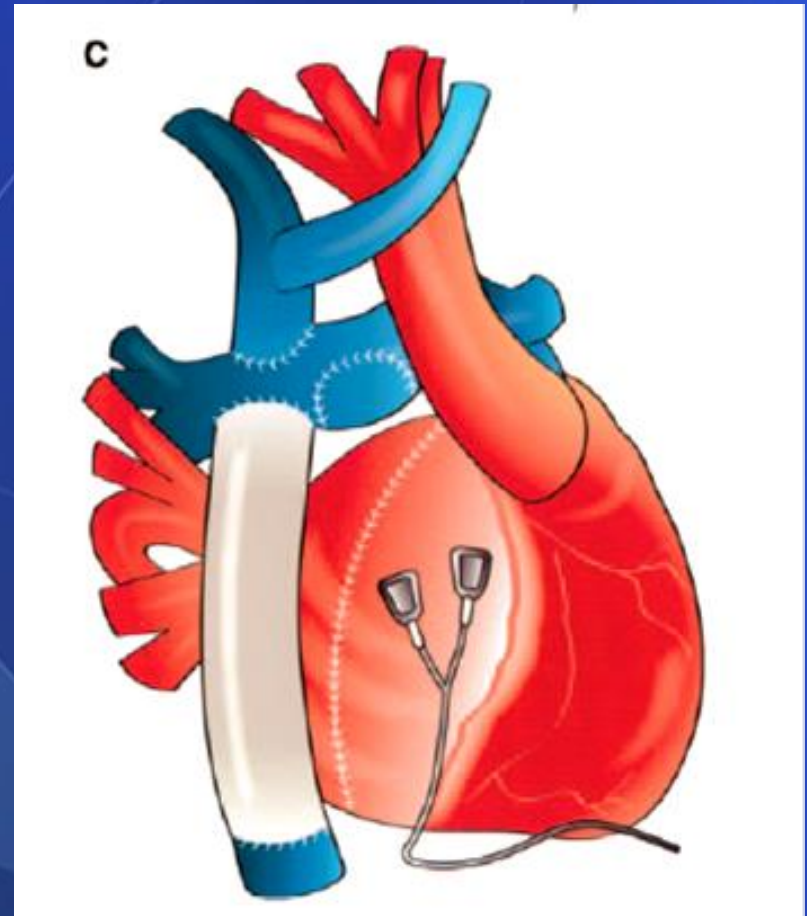
FONTAN

Surgical Techniques

The intracardiac lateral tunnel Fontan



The extracardiac Fontan



Optimal Fontan Physiology

- *Central venous pressure (CVP)* 10-15 mmHg
 - *Pulmonary artery pressure (PAP)* 10-15 mmHg
 - *Left atrial pressure (LAP)* 5-10 mmHg
-

- Transpulmonary blood flow depends on the systemic venous pressure
- Low pulmonary vasc. Resist. (PVR) is crucial
- Transpulmonary gradient (PAP-LAP) is a major determinant of pulmonary blood flow

FONTAN OPERATION

CARDIAC REQUIREMENTS

- Unobstructed ventricular inflow (no AV Valve sten. or regurg.)
- A reasonable ventricular function
- Unobstructed outflow (no Sub. Ao St. no Arterial Hyp. No CoAo)

PULMONARY REQUIREMENTS

- Non-restrictive connection
- Good size pulm. arteries without distortion
- Well developed distal vasc. bed
- Near normal pulm. vasc. resist. (PVR/ <2.5 U/m²)
- Unobstructed pulm. venous return

FONTAN IN ADULT

PVR MAY INCREASE IN THE LONG-TERM FOLLOW UP

Possible mechanisms:

- a) Micro emboli from dilated right atrium**
- b) Aging**
- c) Obstructed airways caused by lymphatic dysfunction**
- d) Lack of pulsatile flow causing a release of endothelium -
derived vasoactive molecules such as “endothelin-1”**

FONTAN FAILURE

Potential risk factors

Long-term chronic hypoxemia

Ventricular volume overload

Arrhythmias

Thromboembolism

FONTAN IN ADULT

Late Fontan Complications 1

- **Worsening cyanosis**
- **Shunting through a baffle leak or residual ASD**
- **Pulmonary vein compression**
- **Systemic venous collateralization**
- **Pulmonary arteriovenous malformation**
- **R to L Shunt**
- **Diaphragmatic paresis**

FONTAN IN ADULT

Late Fontan Complications 2

- Arrhythmias (only 19% pts are arr. free at 20 yrs)
- Heart failure (diastolic dysfunction is common 72%)
- Tromboembolism (occurs in 8-25% of pts)
- Protein losing enteropathy (is present in 3-15% of pts)
- Liver diseases (50% of pts have hepatomegaly)
- Plastic bronchitis (mucous acellular bronchial cast formation)
- Coronary sinus hypertension

FONTAN FAILURE

Solutions

Medical therapy

Fontan Conversion

Assist Devices

Cardiac Transplantation

FONTAN CONVERSION

Surgical revision should be considered in pts with failing Fontan circulation (mortality rates 2.4% - 6.7%)

Surgery typically involves :

- Right atrial debulking
- Conversion to ECC
- Surgical cryoablation to treat R. A. Arr.
- Epicardial dual chamber P.M. implant
- Surgical correction of any anatomical resistance to flow
(Pulm. Branch repair or AV valve repair)
- Removal of thrombus

FONTAN IN ADULTS

San Donato Experience

From December 1998 to February 2013

125 patients operated with Fontan procedure

36 adult patients

Male 22; Female 14

Mean age 28 years (12-43)

FONTAN IN ADULTS

DIAGNOSIS	NUMBER OF PATIENTS
Tricuspid atresia	19
Univentricular heart	10
Double inlet left ventricle	3
Double inlet right ventricle	1
Complex corrected transposition of great arteries	1
Unbalanced A-V canal	2

FONTAN IN ADULTS

Prior Palliative Procedures

PROCEDURES	N
Blalock-Taussig	20
Waterston	2
Pulmonary artery banding	3
Glenn	11
Atrial septostomy	2
Atrial septal defect closure	3
Prior cavopulmonary anastomosis	25

Some patients had more than one procedure

FONTAN IN ADULTS

TOTAL PATIENTS: 36

26 FONTAN CONVERSION	(2 deaths)	7.7%
10 PRIMARY FONTAN	(0 deaths)	0%
TOTAL	36 (2 deaths)	5.5%

FONTAN CONVERSION

N patients	26
Mean age	29,6 years (12-43)
Mean Age at first Fontan	9,8 years (2-29)
Atrio-pulm. connection	23
Atrio-pulm. valved conduit	2
Atrio-ventr. valved conduit	1
Indication for conversion	
Arrhythmia	21
NYHA III-IV/arrhythmia	3
Atrio- pulmonary venous obstr.	2

FONTAN CONVERSION

Surgical technique

Extracardiac conduit	25
Intracardiac conduit	1
Fenestration	2
Maze	18

FONTAN CONVERSION

Early Nonfatal morbidity

COMPLICATIONS	N	%
Prolonged pleural effusion (> 15 days)	6	27
Atrial Arrhythmias	8	36
Permanent pace maker	11	50
Tamponade	1	0.4

FONTAN CONVERSION

FOLLOW UP (24/24)

Mean 83 months (28-161)

NYHA I-II	22
MOD. LIMITATION OF EXCERCISE CAPACITY	1
ARRHYTHMIA	3
PROTEIN-LOSING ENTEROPHATHY	1

FONTAN CONVERSION

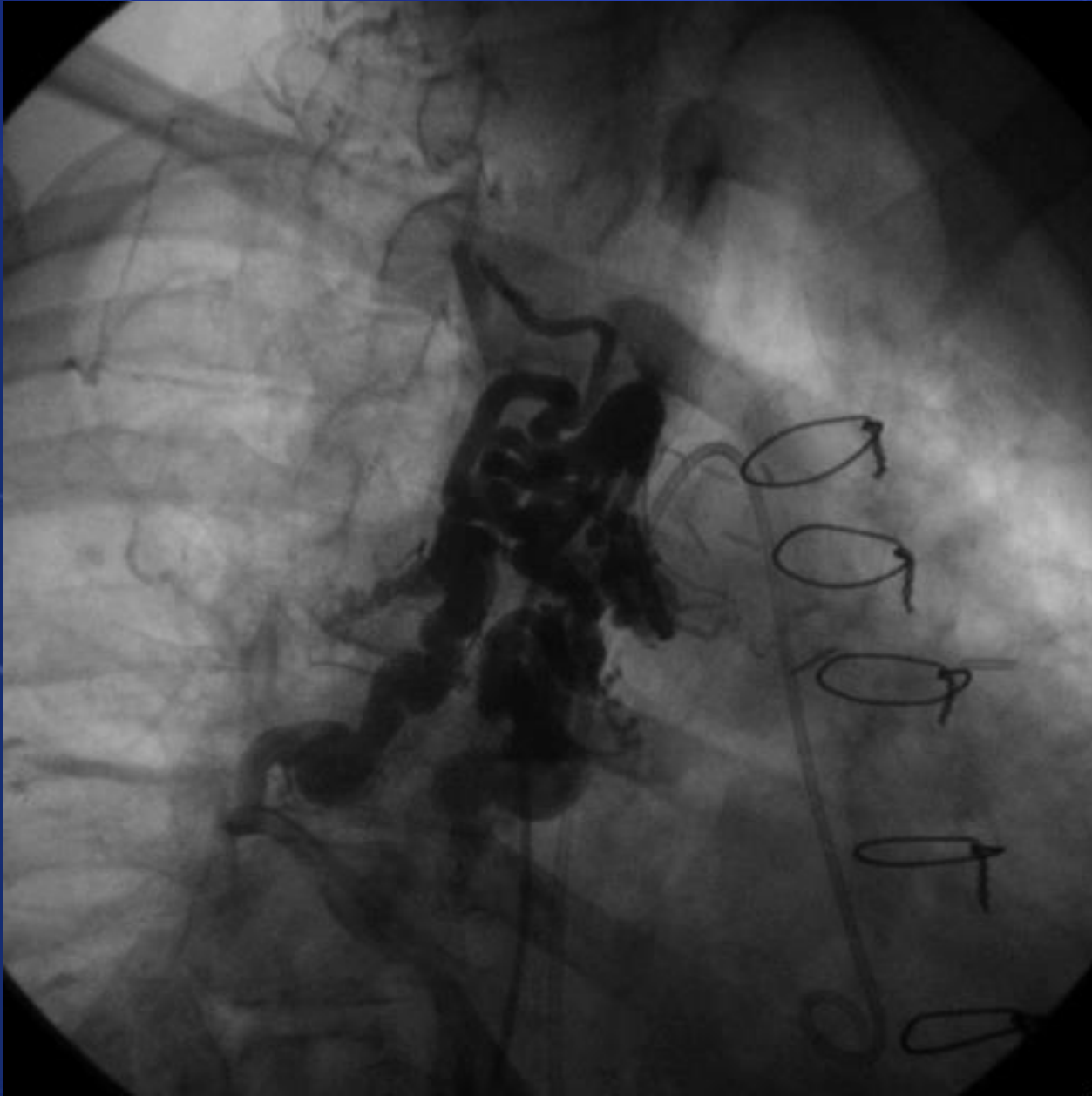
Procedures during follow up

TRANSPLANTATION 1

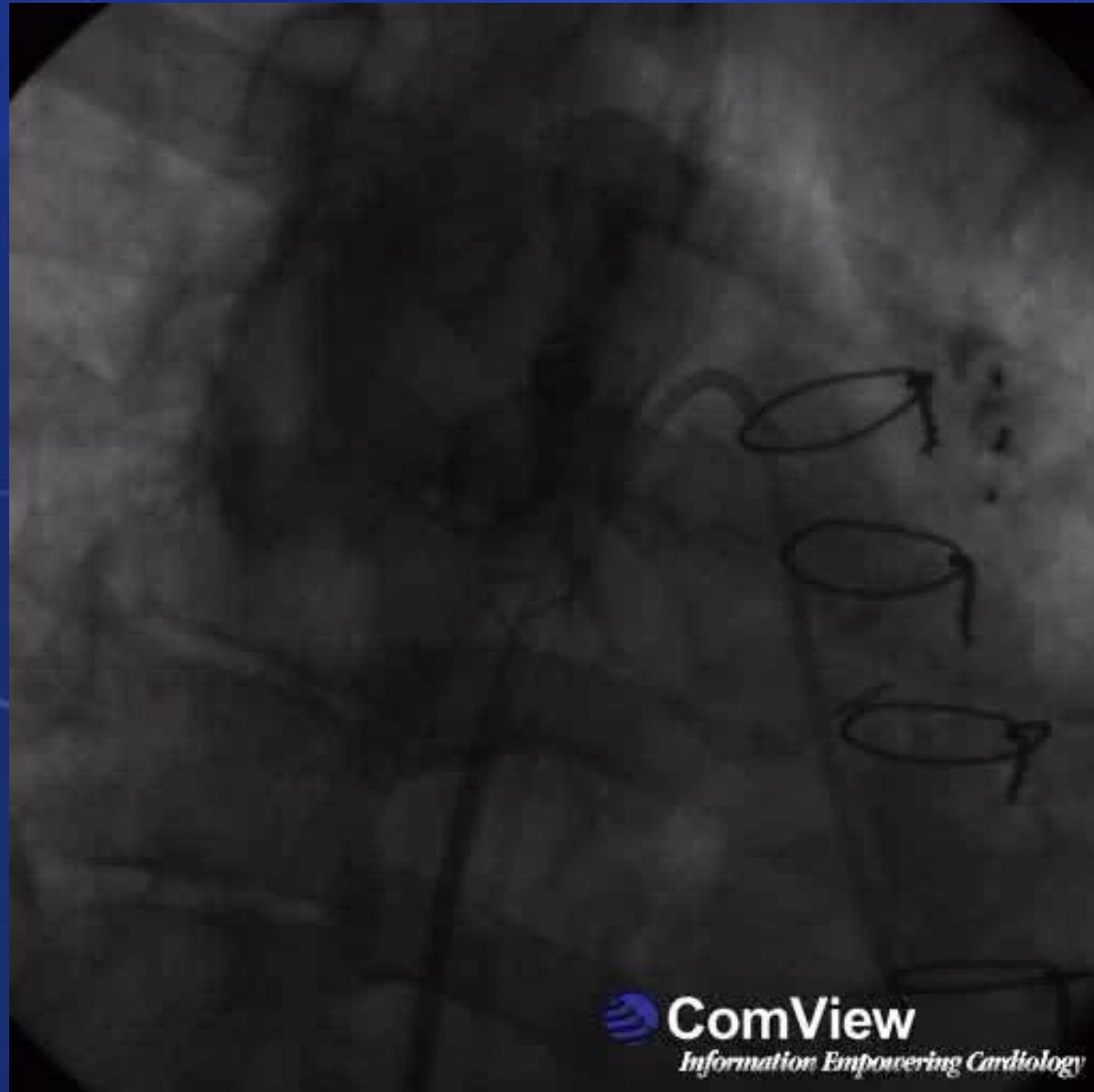
VALVE REPLACEMENT 2

COLLATERAL EMBOLISATION 1

Pulmonary arteriovenous malformation



Pulmonary arteriovenous malformation



PRIMARY FONTAN

N Patients	10
Mean age	28,6 years (17-35)
Indication for Fontan	
Cyanosis	8
NYHA III-IV	2

PRIMARY FONTAN

Early Nonfatal morbidity

COMPLICATION	N	%
Prolonged pleural effusion (> 15 days)	2	20
Atrial Arrhythmias	2	20
Permanent pace maker	2	20
Tamponade	1	10

PRIMARY FONTAN

Surgical technique

Extracardiac conduit	9
Intracardiac conduit	1
Fenestration	1
Maze	2
A-V valve repair	1

PRIMARY FONTAN PROCEDURE

FOLLOW UP (10/10) Mean 97.8 months (72-133)

NYHA I-II	8
MOD. LIMITATION OF EXCERCISE CAPACITY	1 (30 y)
SEV. LIMITATION OF EXCERCISE CAPACITY	1 (43 y)
ARRHYTHMIA	1
RENAL	1
CEREBRAL VASCULAR ACCIDENT (TIA)	1

FONTAN PROCEDURE

ROUTINE HEALTH CARE OF THE FONTAN PTS

- Adult Fontan pts should be managed by a cardiologist with an expertise in adult congenital heart disease
- Annual visit involving pts history, physical examination - ECG - Chest X-ray and echocardiography
- Prophylactic use of aspirin
- Anticoagulation with warfarin is recommended for all pts with atrial arrhythmias-thrombus-atrial level shunting or previous embolic event
- ACE inhibitors should be used in pts with ventricular dysfunction

Literature review

- **Early mortality:** 8.3%
- **Early complications:**
 - Prolonged pleural effusion 30%
 - Arrhythmias 21%
- **Freedom from late mortality:** 89% at 5 y, 75% at 10 y, 68% at 15 y
- **Significant risk factors:** mean PAP >15 mmHg, male gender, age > 30 y
- **Freedom from late reoperation:** 89% at 5 y, 85% at 10 y, 81% at 15 y
- **Late complications:** arrhythmia 42%; pacemaker 15%

FONTAN

Conclusions

Longterm results in pts with a low risk profile show good overall results

In less than ideal pts important sequelae persist

FONTAN FAILURE

Lung function and aerobic capacity



Greatly diminished values

Pts with an early surgical procedure obtained
higher values of $\dot{V}O_2$ max

FONTAN IN ADULTS

Epidemiology

- Univentricular heart
 - 54 cases per million live births
- Hypoplastic left H-S 2-3 x10.000
- Tricuspid atresia <1 x 10.000
- DILV 1% all congenital heart malformation

FONTAN IN ADULTS

Original Fontan Criteria

- Age ≥ 4 years to < 15 years
- Normal sinus rhythm
- Normal systemic venous return
- Normal right atrial volume
- Mean pulm. art.pressure ≤ 15 mmHg
- Pulmonary arteriolar resistance < 4 WU/m²
- Pulmonary artery to aortic diam. ratio ≥ 0.75
- Left ventricular EF ≥ 60
- Competent mitral valve
- Absence of pulmonary artery distortion

FONTAN IN ADULTS

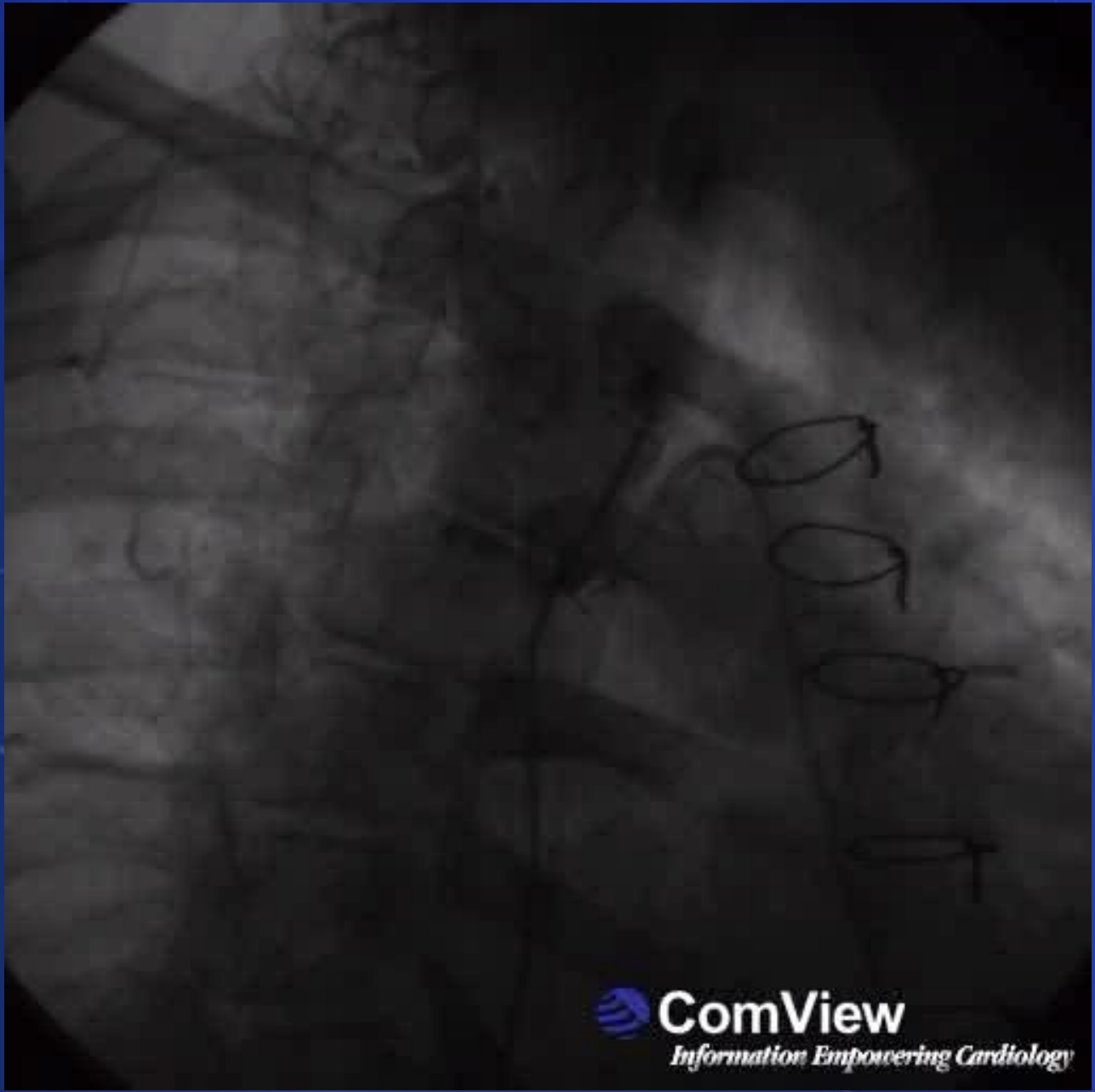
Complications

- Arrhythmias
- Ventricular dysfunction
- Valve regurgitation
- Vascular
- Coagulation disorders
- Protein losing enteropathy (PLE)
- Liver dysfunction
- Neurological
- Plastic Bronchitis

FONTAN IN ADULTS

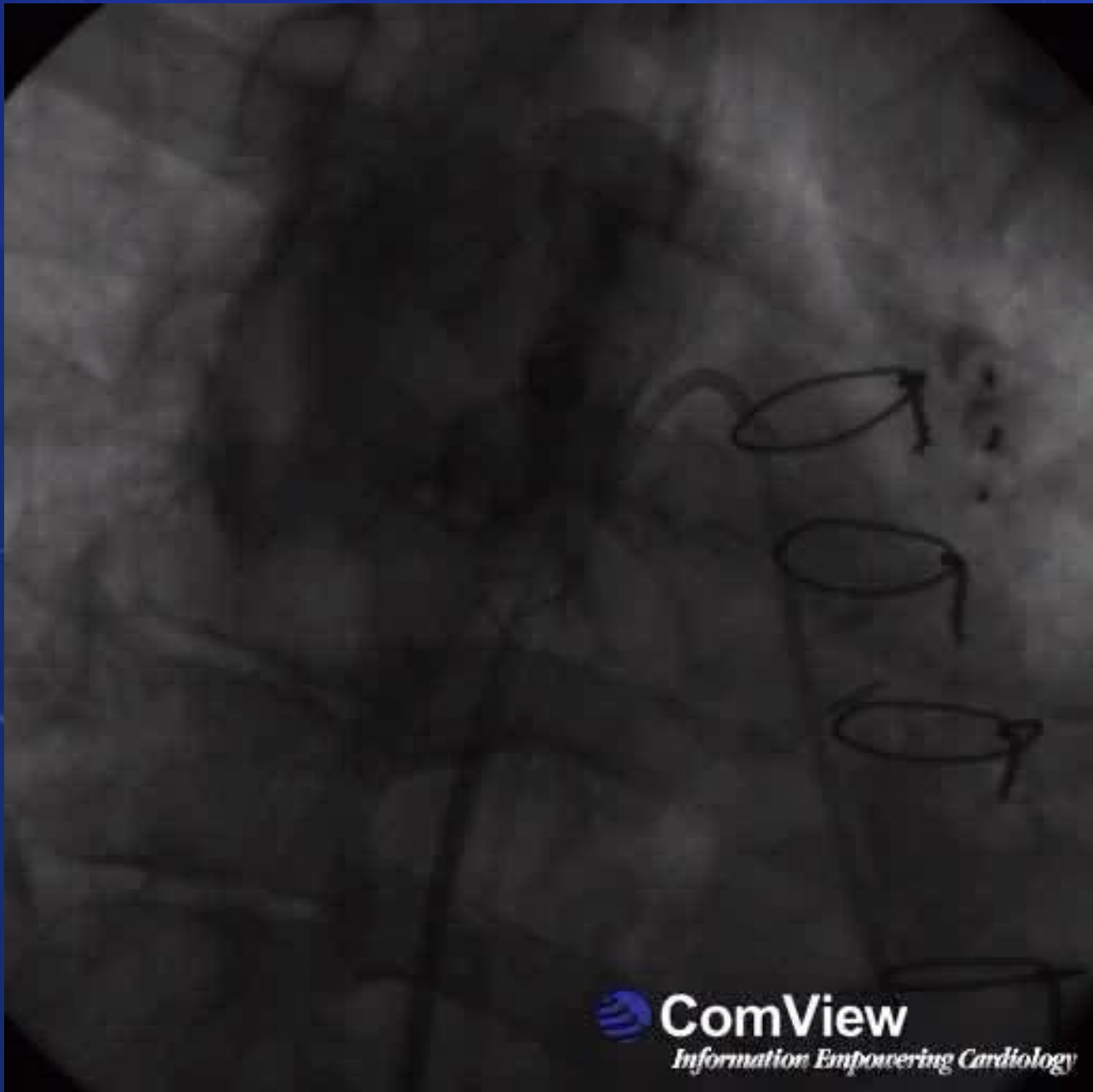
Univentricular Heart

- Absence of 1 AV valve (mitral or tricuspid atresia)
- Hypoplastic left heart syndrome
- Double inlet left (DILV)
- Common AV Valve and only 1 well developed ventricle
- Only 1 well developed ventricle and heterotaxy syndrome



ComView

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