



## ROLE OF THE FONTAN OPERATION FOR TREATMENT OF SINGLE VENTRICLE HEART DISEASE AT CHO RAY HOSPITAL

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## INTRODUCTION

- Functional single ventricle disease is rare, accounts for 4-8/1000 newborn and 7.7/100 of congenital heart disease.
- Poor prognosis without surgical treatment (75% die within the first 3 years of life.)
- Fontan surgical the purpose of passively diverting all blood from the systemic vein to the lung helps repairing this pathophysiology for normal achivement.



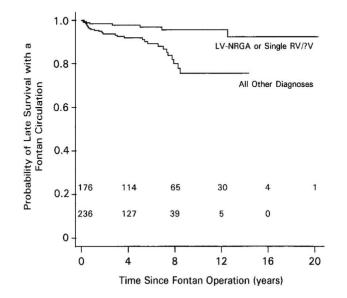


Fig. 5. Kaplan-Meier estimates of the probability of late survival with a Fontan circulation as a function of diagnosis. Time zero is 30 days after the Fontan operation. *LV-NRGA*, Left ventricle with normally related great vessels; *Single RV/?V*, single right ventricle or ventricle with unknown morphology.

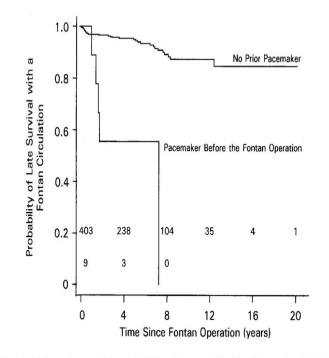


Fig. 6. Kaplan-Meier estimates of the probability of late survival with a Fontan circulation in patients with and without a pacemaker before the Fontan operation. Time zero is 30 days after the Fontan operation.



## INTRODUCTION

- Techniques of Fontan operation have many improvements over time, helping increasingly optimize treatment results.
- The Fontan operation by tube graft with fenestation which made in many coutries initially has good results at Cho Ray hospital.



## PATHOHYSIOLOGY OF FUNCTIONAL SINGLE VENTRICLE

#### Anatomy(1)

- Jacobs and Anderson : Anatomical characteristic with one or two ventricles chamber but only one ventricle has normal function.
- The remaining ventricle which is non- existent or hypoplastic can not resonsible for pulmonary or systemic function.

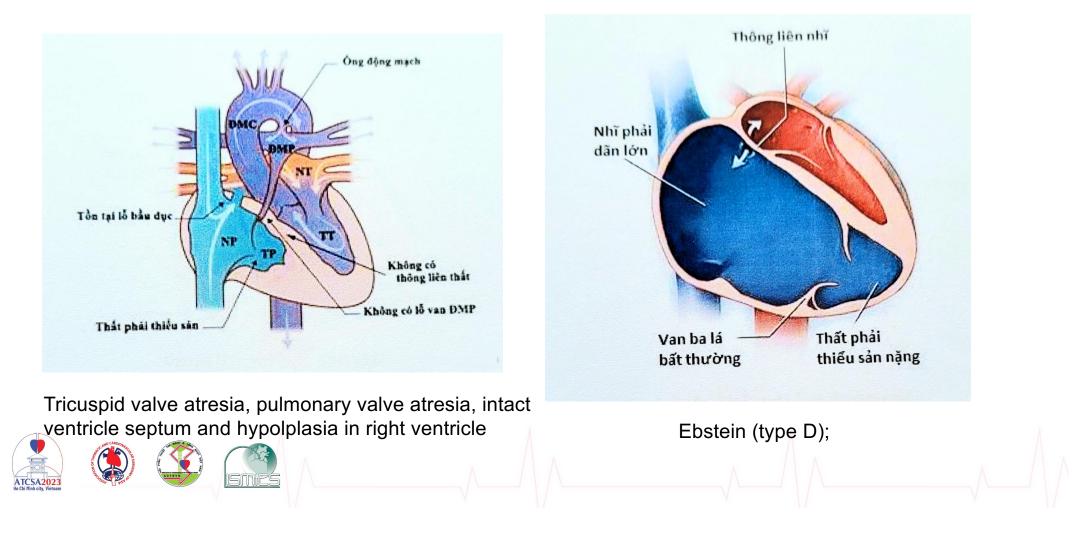
## PATHOHYSIOLOGY OF FUNCTIONAL SINGLE VENTRICLE

#### Anatomy (2)

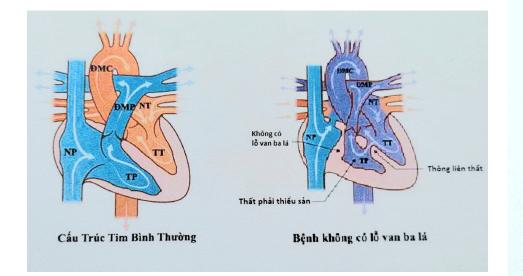
Some single ventricle diseases: Tricuspid atresia, Pulmonay valvular atresia with intact ventricular septum and hypoplasia RV, Ebstein type D, common double right or left ventricle inlet; mitral valve atresia in HHLS, unbalance A-V canal,,,,



## SINGLE VENTRICLE DISEASES

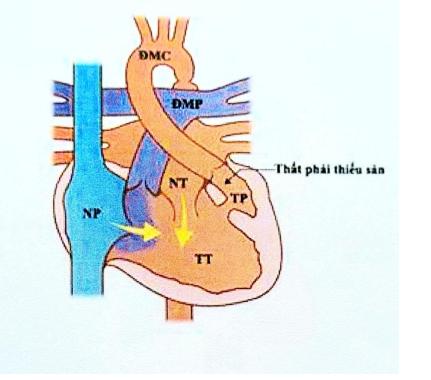


## SINGLE VENTRICLE DISEASE



Common double right or left ventricle inlet; mitral valve atresia in HHLS

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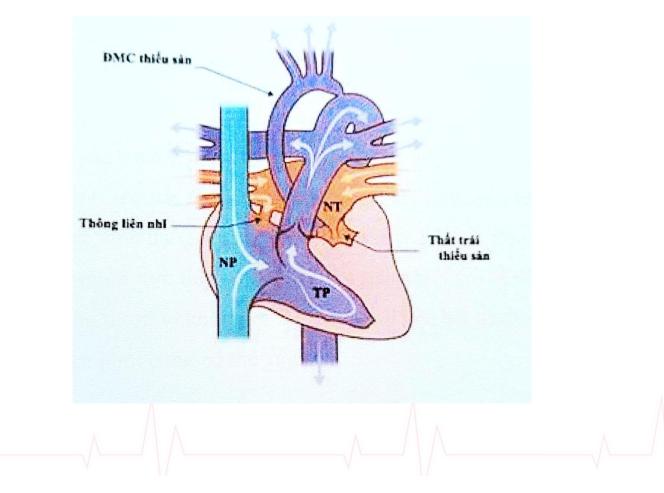


Hypoplasia of RV

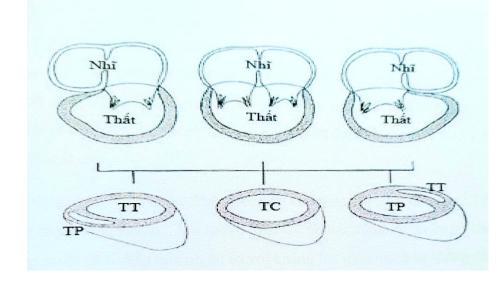
## SINGLE VENTRICLE DISEASES

Mitral valve atresia in HHLS

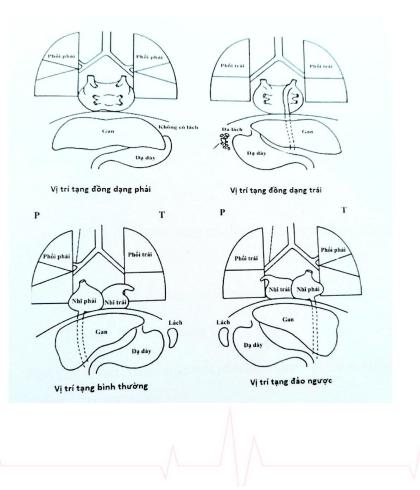
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# SINGLE VENTRICLE DISEASES



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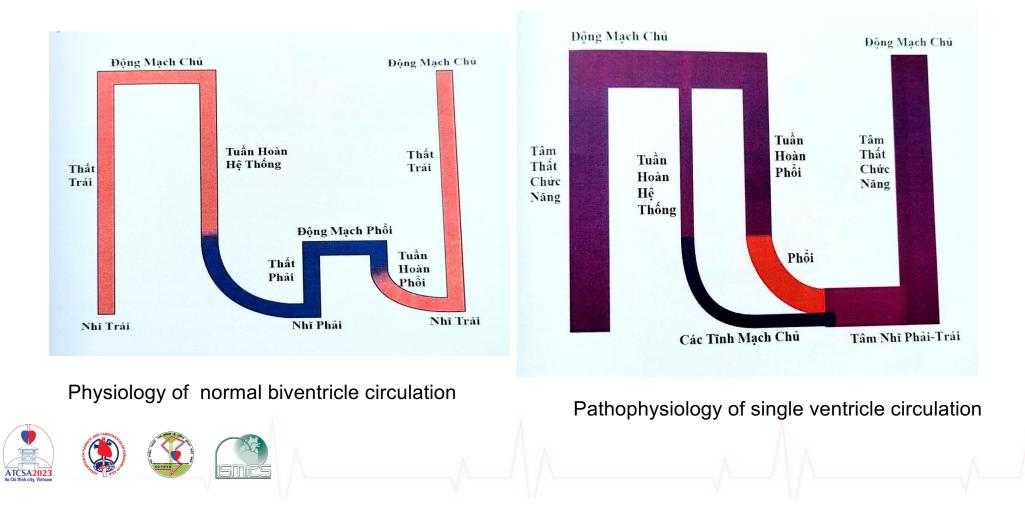
## PATHOPHYSIOLY OF SINGLE VENTRICLE

#### The functional classifications:

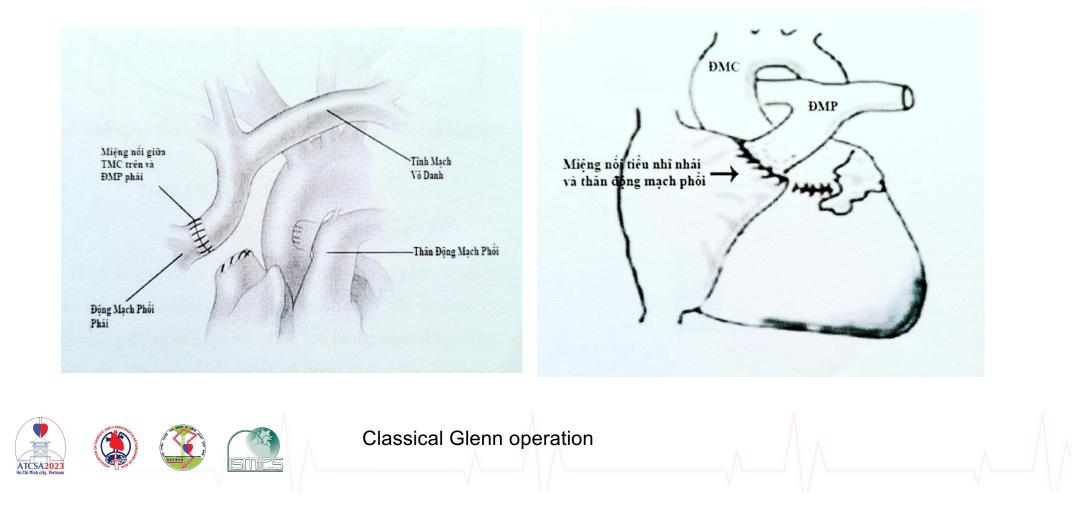
- The dominant ventricle as left ventricle structure + hypoplastic rudimentary right ventricle without function.
- The dominant ventricle as right ventricle structure + hypoplastic rudimentary left ventricle without function.
- The dominant ventricle can not decả hai thất.

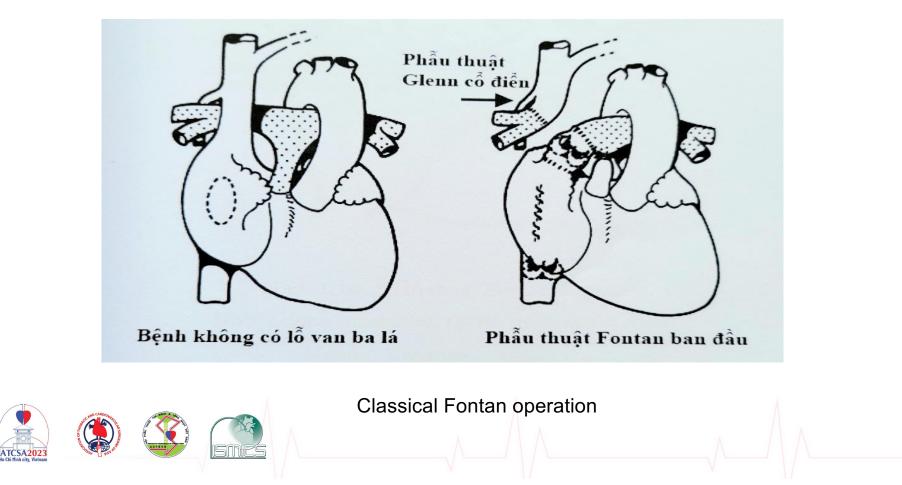


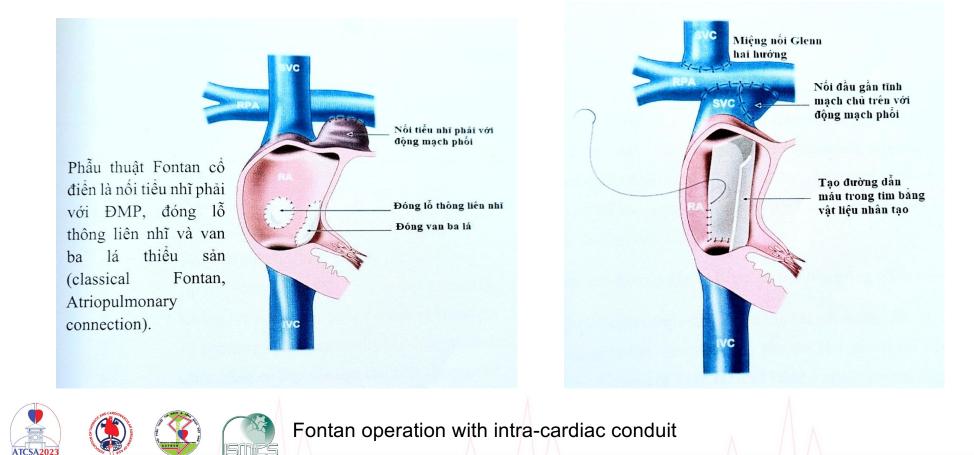
#### PATHOPHYSIOLY OF SINGLE VENTRICLE



#### **PREPAIR FOR FONTAN OPERATION**









- Currenly, development of techniques and improvement in resuscitation
- Fontan operation applied by extra cardiac with graft tube+ fenestration
- 1990 : Marcelletti and Co, created extra cardiac blood path from the IVC to the PA with Dacron or Gortex tube in patients whose

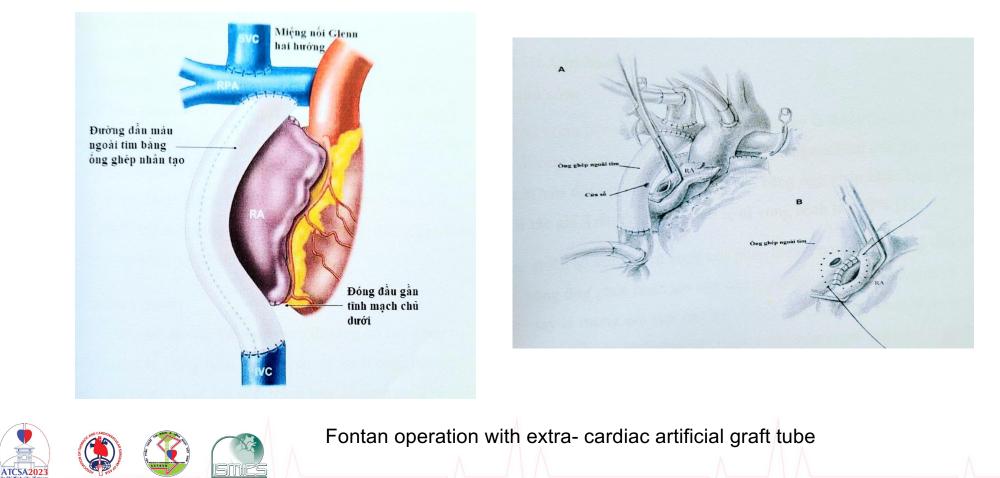
underwent bidirectional Glenn surgery.



#### Advantages:

sutures

- Feasibly perform in patient with single ventricle anatomy
- Heart beating with CPB (prevent the side effects from CPB and cardioplegia fluid)
- Uniformity in size between the graft and diameter of IVC (prevent narrow the path and good control the blood flow to the lung)
- Reduced risk of arrhythmia due to stress in atrial wall or atrial



## DIAGNOSIS

Echocardiology

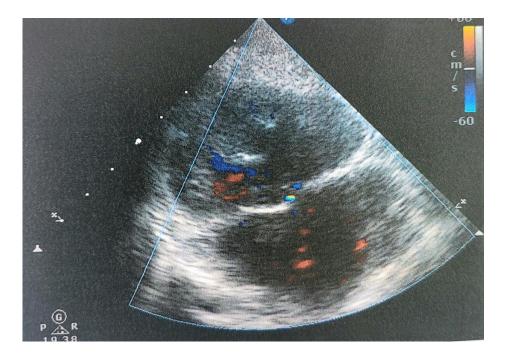
Thoracic MS CT

Cath-lab diagnosis



## DIAGNOSIS

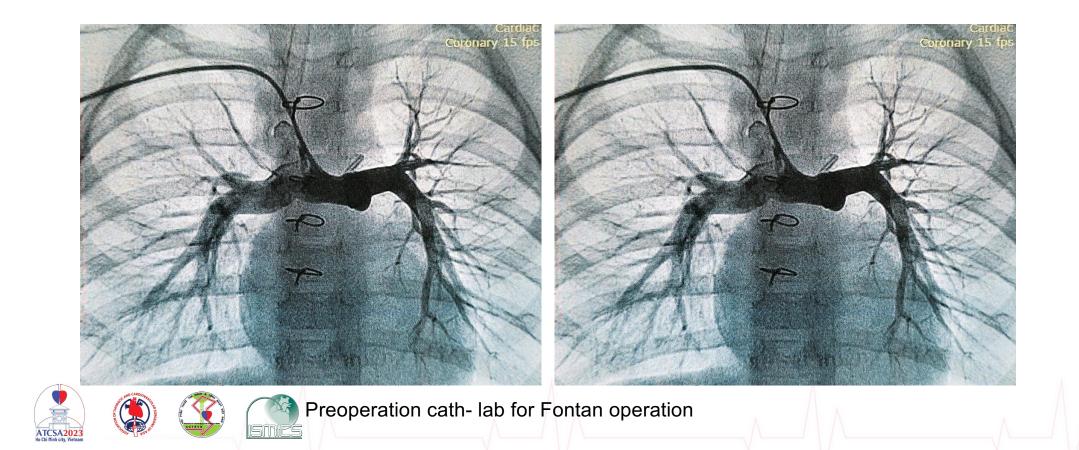






Echo cardiology preoperation

## DIAGNOSIS



## OBJECTIVE

Evaluating early results of Fontan operation using the extra cardiac conduit with fenestration in the treatment of functional single ventricle heart disease at Cho Ray hospital

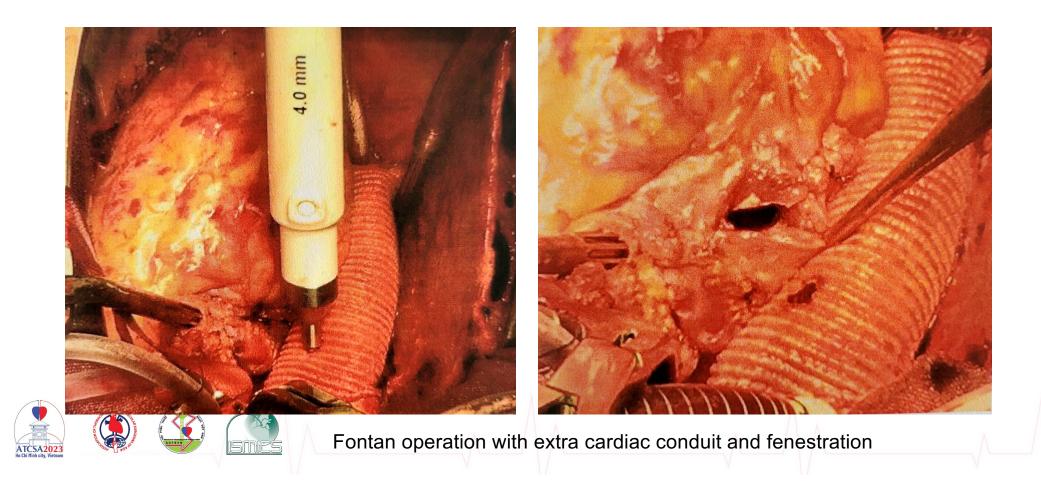


# MATERIAL AND METHOS

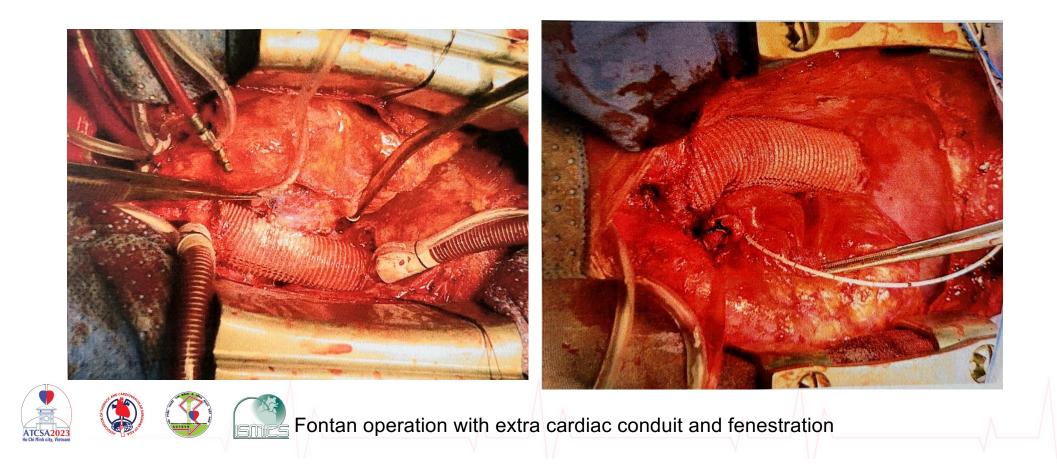
- Retrospective study
- Duration: from 1/2017 to 12/2021
- Subjects: 31 patients with functional univentricle heart disease who underwent Fontan operation using the extra cardiac conduit with fenestration.



## **TECHNIQUES**



## TECHNIQUES



RESULTS

#### Patient characteristic (n=31 pts)

Male: 18 pts (59,2%)

Do Anh Tien et al (2019):

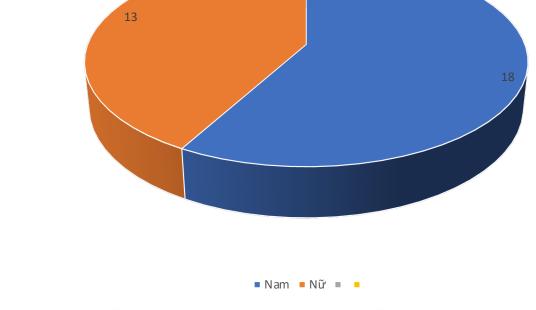
male/ female: 35/26

Kotani et al (2018):

303/197 (2/1)

484/289 (2/1)

Tacy Downing et al (2017):



Gendre



#### **Patients characteristics**

- Mean age:  $66,3 \pm 11,4$  months (19 204)
- Ohye et al(2016) : Fontan operation was perform from 18 months to 4 years old
- Masamichi Ono et al. (2019): mean 2,3 (1,8 3,8)
- Edward Malec et al (2017): 3,7 (1,1 6,3)
- Đỗ Anh Tiến et al (2019): 5,6 (2-14)

# Historical treatment

Historical treatment	Number of pts	%
PDA Stent	5	16,1
B-T shunt	8	25,8
PA banding	2	6,45
Bidirectional Glenn procedure	31	100

Mean Glenn operation duration : 15,1 ± 3,4 months



# Clinical signs NYHA classification

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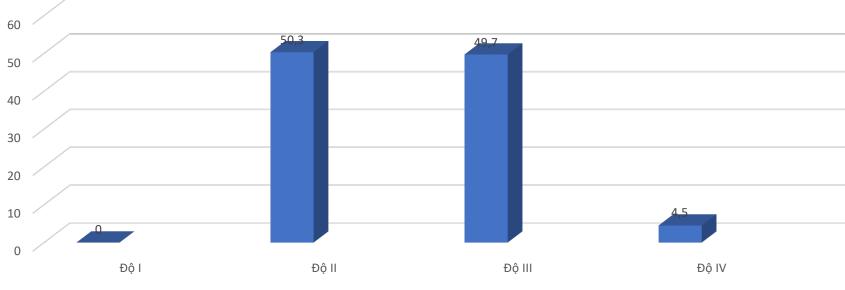


Chart Title

■ Suy tim NYHA ■ Column2 ■ Column1

Most of patients had NYHA grade I: 0%; II: 50,3 (%). Pts

49,7(%) grade III, no patients with grade IV.

## Clinical and para clinical signs(n=31)

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Value	Mean	Range
SpO2 (%)	80,2 ± 3,4	78-92
Heart rate (beat/minute)	90,3 ± 15,6	76-110
Systolic arterial pressure (mmHg)	95,2 ± 9,8	87-138
Diastolic arterial pressure (mmHg)	59,1 ± 6,3	50-83
Cardio-thoraci ratio	0,56 ± 0,02	0,45- 0,6
Hct (%)	54 ± 7,26	50-67
EF (%)	61,4 ± 7,3	52-78

Most of patients had signs of central cyanosis and sinus rythm. EF > 60% account in 69% pts.

## Cath-lab diagnosis

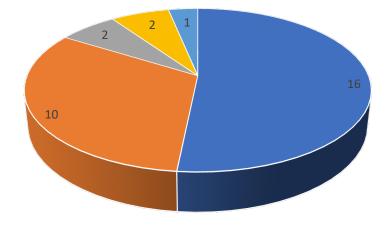
Value	Mean	Range
Nakata index (mm2/m2)	245 ± 80,7	220 - 312
Mean PA pressure (mmHg)	11,4 ± 2,3	8 - 15
PVR (wood/m2)	$1,8 \pm 0,9$	0,9 – 2,2

Most of patients had Nakata index > 150 (mm2/m2) and mean PA pressure <15 mmHg with low PVR pre operation



# Morphometric of cardiac

Tricuspid atresia accounts high prevalent as 51,6%; total A-V canal unbalance + PA hypoplastic or atresia



Không lỗ van ba lá
Kệnh AV - hẹp ĐMP
DORV - thiểu sản thất T = PS- thiểu sản thất phải
DOLV - thiểu sản thất P

Mineto Kamata et al (2017): right side univetricle accounts as 51,4%; left- side :27,8%; indefinite : 20,8%

#### Surgical characteristics

Characteristic	Number of pts	%
A-V valve repair	2	6,45
Enlarment in PA branches	4	12,9
Conduit fenestration	31	100
Size of Conduit 20mm	22	72
size of conduit 18mm	9	28

Elizabeth H. Stephen et al (2019): AV repair by Alan Carpentier Yasuharu et al (1997): A-V annular plasty in dilate annulus AV valve



#### Surgical characteristics

- Koichi Sughimoto et al (2017): the survival rate in patients with Fontan operation after 3 years was 66%.
- Keiichi Itatani et al (2009) the size of graft tube with 16 18mm suitable for pts 2 – 3 years old



#### Surgical features:

- Scott et al (2006): child patients with 15 kg can use the conduit
   20mm for good flow as adult patients
- Brakel et al (2014) using the diameter of conduit < 16mm high risk of narrow.
- Conduit with size as 18 20mm are optimally effective even for children less than 12 kg.

## Surgical features

- Alan G. Magee et al(1998): divided helical group of the PA as 4 group in PA plasty with Fontan operation.
- Igor E. Konstantinov et al (2016): using the patch to plasty the PA branches is better than cutting the graft for widen the connetion to the PA



#### Surgical feature

- Qiang Fan et al (2017):Using the Goretex artifical tube with circle cover make fenestration may difficult in operation
- Pan et al (2016): Shoud make fenestration in condiuts for high risk patient



## Surgical features

Value	Mean	Range
CPB time (minutes)	83,9 ± 28,4	67 - 158
Aortic clamp time(minutes ) (n=5)	60,3 ± 11,5	45 - 101
PA pressure right after CPB stop (mmHg)	15,3 ± 1,42	10 - 17
SpO2 (%)	90,4 ± 5,16	88.95
Heart rate ( beat /minute)	$101,4 \pm 4,3$	78 - 120
Systolic arterial pressure (mmHg)	99,2 ± 13,8	86 - 132
Diastolic arterial pressure (mmHg)	60,3 ± 4,5	55 - 78

 $D\tilde{0}$  Anh Tiến et al (2019) mean CPB time 98 ± 31,15 minutes Sarkar et al (2017): 110,2 ± 1,3 minutes; long time in CPB make more side effects in inflamtory or respiration function.

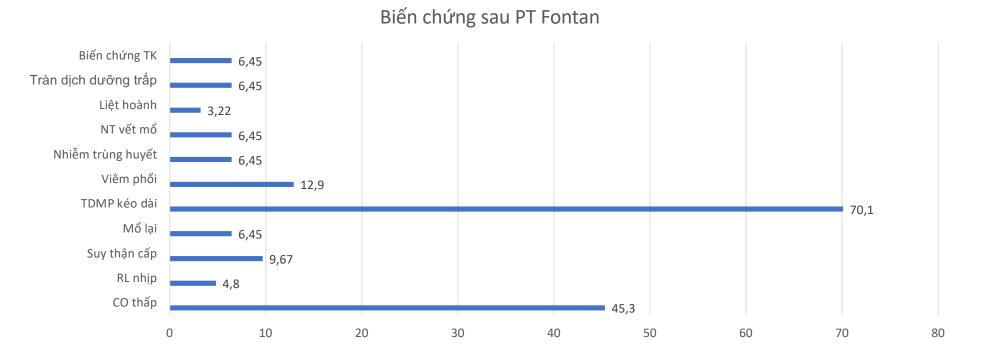
#### THE RESULTS Post operative features

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VALUE	MEAN	RANGE
Machanical ventilator mean time( hours)	50,2 ± 103,7	5 - 201
CCU mean time ( days)	7,6 ± 9,5	2 - 23
Post operative mean time(days)	20,7 ± 15	10 - 78
Mean Time of pulmonary effusion drain (days)	11 ± 24,3	2 - 54
The fluidd volume per day (ml)	250,6 ± 102	103 - 1305
SpO2 post operation (%)	92 ± 3,55	90 - 96
Mean Hct post. operation(%)	$40,1 \pm 1,4$	38 - 46

Mechanical ventilator time under. 2 days ; Post operative time was quite long. The pulmonary effusion drainage has alot

#### Post operative complications



Gupta et al: lung effusion drain after Fontan operation account for 13 – 39%; Minh Nhựt et al found in 39%. High prevalent of heart failure appeared after Fontan operation



#### Hospital mortality

• Hospital mortality rate : 3 BN (9,67%)

Etiology: 02 pts with low cardiac output and multi organs failure

01 pt with blood infection and shock sepsis

- Marcelletti et al (1999): hospital mortality rate 10%;
- Trần Đắc Đại et al (2020): with 8,97% death rate.

# Short term results mean following time : $17,3 \pm 34,1$ (month) NYHA classification pre and post operation

NYHA	Pre ope	1 month later	6 months later	1 year later
I	5	8	17	21
Ш	18	22	12	9
111	8	1	2	1
IV	0	0	0	0



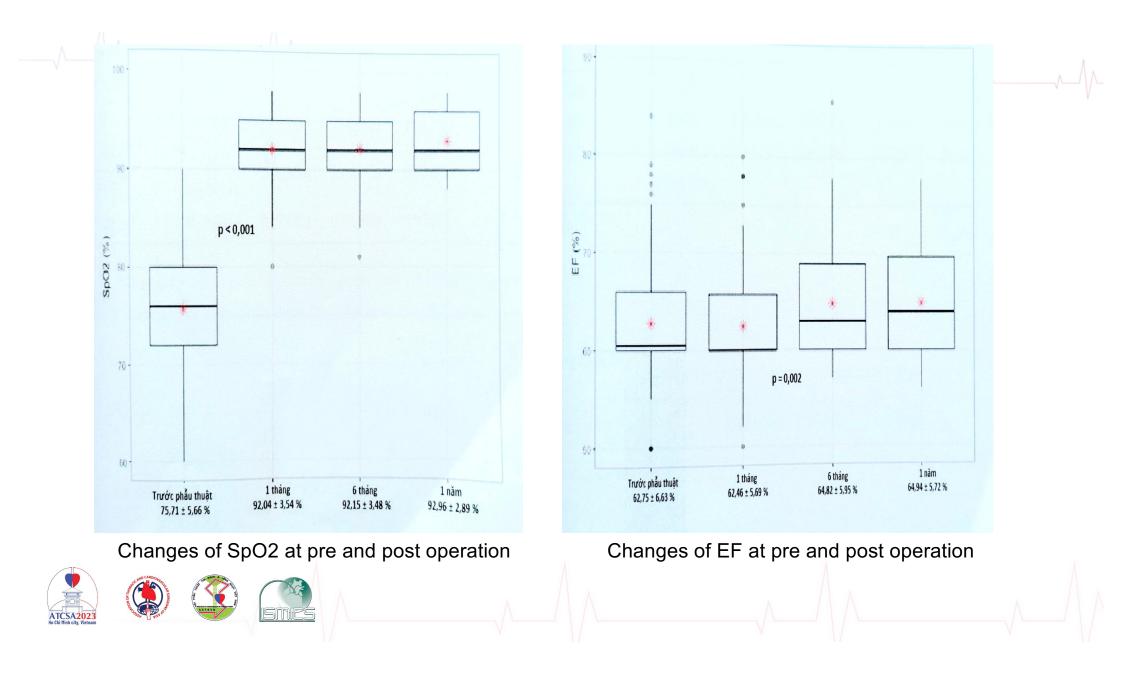
#### The changes of EF (by Echocardiology))

year.

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EF (%)	Value
Pre operation	61,5± 3,7
1 month later	62,37 ± 1,02
6 months later	64,8 ± 5,15
1 year later	64,96 ± 5,12

There is an improvement in EF pre and. After Fontan operation at ;1, 6 months and 1



#### The changes of SpO2 pre vs post operation

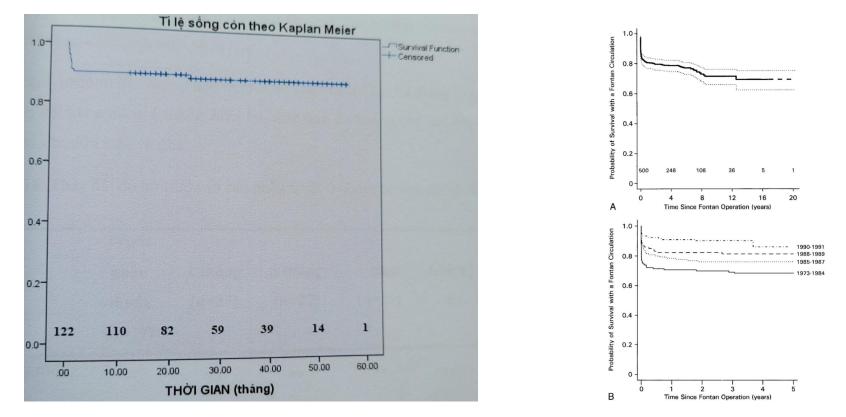
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SpO2 (%)	Value
Pre operation	76,7 ± 6,3
1 month later	92,3 ± 4,51
6 months later	92,8 ± 3,47
1 year	93 ± 1,02

There is a significant improvement changes in SpO2 pre operation vs 1, 6 months and 1 year.

#### The survival rate

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The survival rate in patients with Fontan operation is : 90,2 % at 1 month; 87,3 % at 6 months

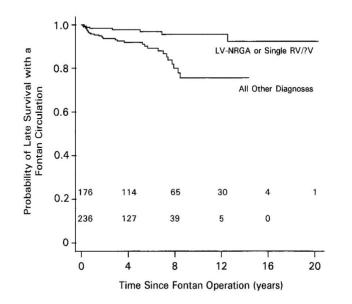


Fig. 5. Kaplan-Meier estimates of the probability of late survival with a Fontan circulation as a function of diagnosis. Time zero is 30 days after the Fontan operation. *LV-NRGA*, Left ventricle with normally related great vessels; *Single RV/?V*, single right ventricle or ventricle with unknown morphology.

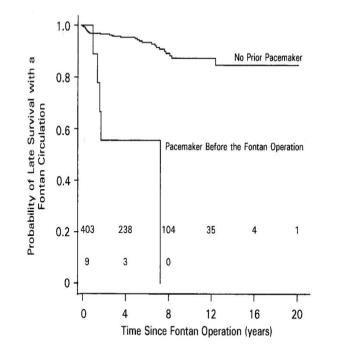
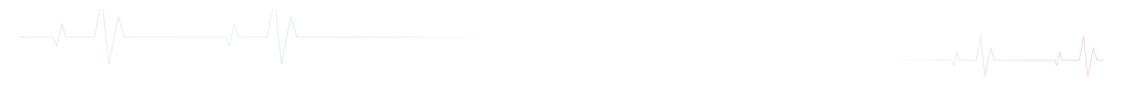


Fig. 6. Kaplan-Meier estimates of the probability of late survival with a Fontan circulation in patients with and without a pacemaker before the Fontan operation. Time zero is 30 days after the Fontan operation.





- Sainz et al :Fontan operation gets good duration exercise (more than 7METs) according to Bruce criteria
- Kreutzer et al : female patients with Fontan operation can get pregnant and have a normal baby



- Yves et al : Fontan operation with extra cardiac tube help reduce the arrythmia
- Minh Nhựt et al : the survival rate after Fontan operation is 90,2 % at 6 months ; Marcelletti et al is 90%. Edward et al found the early survival rate as 100%.



# CONCLUSION

- Study in 31 patients with functional single ventricle heart disease were underwent. Extra cardiac Fontan operation with artificial graft tube with fenestration at Cho ray hospital has good results
- There is low in hospital mortality rate; and low prevalent of post operative complications
- The early survival rate after Fontan by that way is good.

### CONCLUSION

- There is an improvement in cardiac function after Fontan operation
- Recomendation in Fontan operation with extra cardiac artificial tube plus fenestration in most patients get a good and long life







# **THANK YOU VERY MUCH**