



# PERIOPERATIVE EVALUATION AND MID-TERM OUTCOMES AFTER SURGICAL CORRECTION OF ALCAPA IN CHILDREN

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# NỘI DUNG



Thầy thuốc tận tâm - Chăm sóc đất nước

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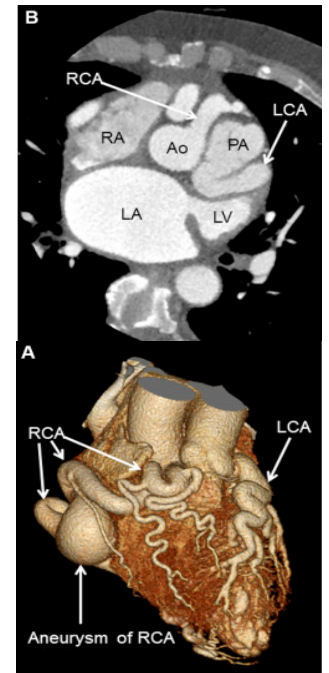


# INTRODUCTION

**ALCAPA** (The anomalous origin of the left coronary artery from the pulmonary artery)

## ALCAPA

- ✓ rare, 0.25-0.5%/CHD
- ✓ one of the most common causes of myocardial ischemia and infarction in children.
- ✓ “coronary steal” phenomenon, left-to-right shunt leads to abnormal left ventricular perfusion → severe myocardial ischemia, global left ventricular dysfunction, various mitral regurgitation, ventricular arrhythmias, sudden death.

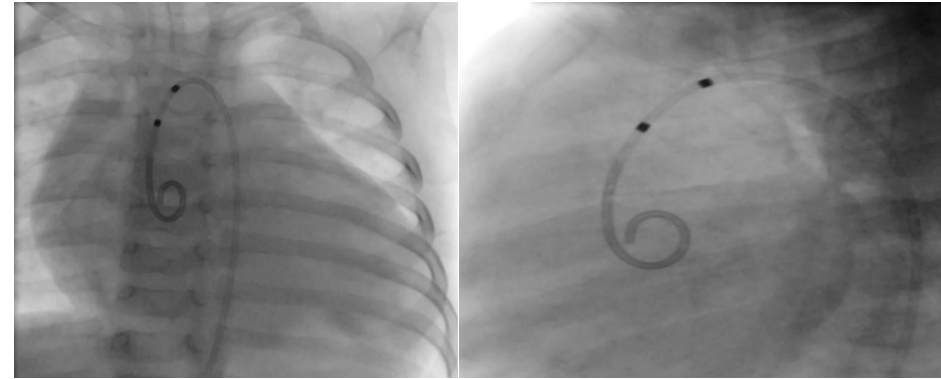


# INTRODUCTION ALCAPA

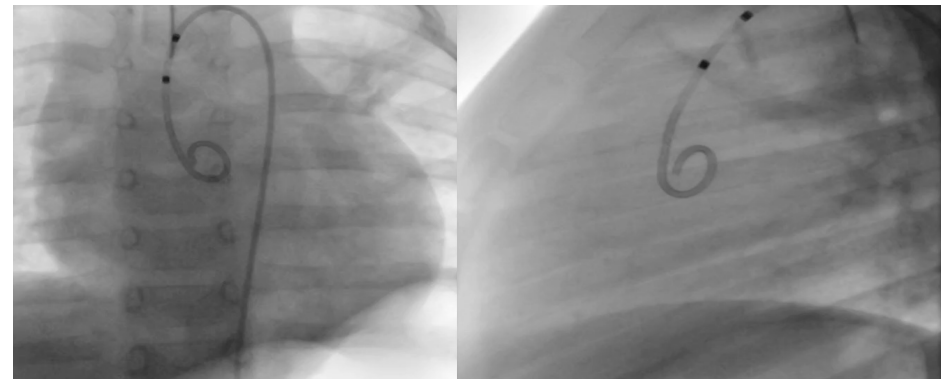


✓ **Infant type:** poor collaterals between RCA to LCA, *90% die before 1 year of age* in untreated patients.

✓ **Adult type:** Abundant collaterals circulation, *90% die in the following years* due to ventricular arrhythmias, ischemic cardiomyopathy.



*Infant type*



*Adult type*



# INTRODUCTION DIAGNOSIS



- Early diagnosis of ALCAPA remains a challenge because the patients has the lack of specificity in the clinical manifestations, which range from asymptomatic to severe myocardial ischemia, heart failure, and sudden death.
- Coordinate diagnostic imaging multimodalities such as ECG, echocardiography, cardiac CTA, CMR, DSA to confirm the diagnosis, help delineate the anatomy and origin of the anomalous coronary.
- Multiple techniques have been introduced to establish a dual-coronary artery system, including
  - ✓ direct reimplantation of the anomalous coronary artery into the aorta (DIACA).
  - ✓ coronary artery bypass grafting (CABG)
  - ✓ coronary baffling procedures



# INTRODUCTION

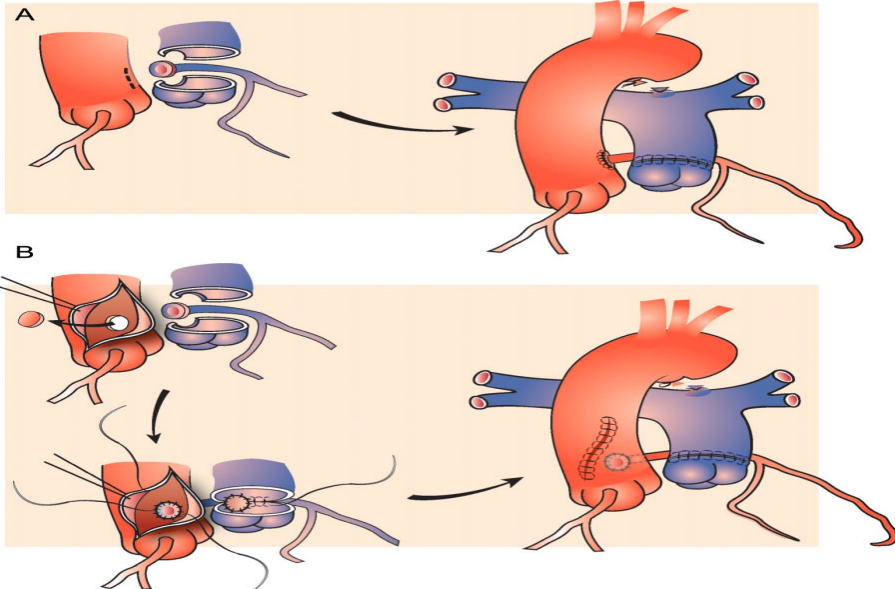
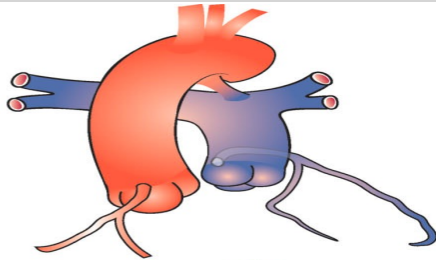
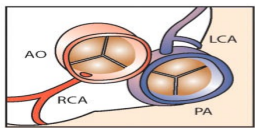
## SURGICAL OPTIONS

Type of Repair	Description	INDICATIONS
<p><b>Direct reimplantation of the anomalous coronary artery into the aorta (DIACA)</b></p>	<p><b>Direct reimplantation of the anomalous artery into the aorta by transferring it with a button of the MPA</b></p>	<p><b>Used in infants; the most anatomic correction and yields excellent long-term results</b></p>
<p>Takeuchi procedure (transpulmonary baffling)</p>	<p>A baffle made from the PA wall is used to tunnel the coronary artery through the MPA into the left coronary ostium</p>	<p>Used in infants when coronary button transfer is not feasible due to unfavorable coronary anatomy</p>
<p>Coronary artery bypass grafting (CABG) combined with ligation of the anomalous LCA</p>	<p>A venous or arterial bypass graft is placed from the aorta to the proximal LAD artery, and the anomalous LCA is ligated at its pulmonary origin</p>	<p>Preferred technique in adults</p>



# INTRODUCTION

## Type of Repair

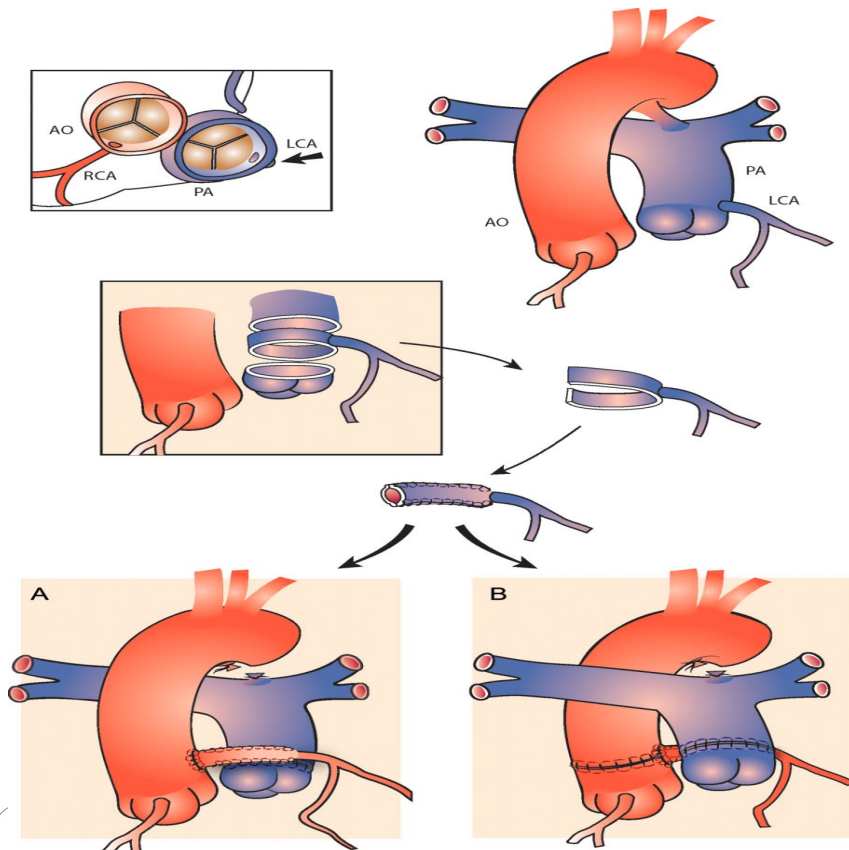


**Surgical reimplantation of ALCAPA from the right facing sinus of Valsalva.**

- **A, Direct implantation into the ascending aorta.**
- **B, Direct implantation under direct vision via anterior aortotomy.**
- AO indicates aorta
- PA, pulmonary artery
- LCA, left coronary artery
- RCA, right coronary artery
- ALCAPA, anomalous left coronary artery from pulmonary artery.

# INTRODUCTION

## Type of Repair



### Surgical reimplantation techniques of ALCAPA from nonfacing sinus of Valsalva.

A, Anterior direct reimplantation by creating cylindrical extension of the coronary artery.

B, Posterior direct reimplantation by creating cylindrical extension of the coronary artery and anterior translocation of PA (LeCompte maneuver) to avoid posterior compression of the implanted coronary artery.

AO indicates aorta  
 PA, pulmonary artery  
 LCA, left coronary artery  
 RCA, right coronary artery  
 ALCAPA, anomalous left coronary artery from pulmonary artery.



# OBJECTIVE

- This study aimed to describe the perioperative clinical features, ECG and transthoracic echocardiographic (TTE) presentations, surgical options and mid-term outcomes after surgical correction of ALCAPA in children at Children's Hospital 1 HCMC.

# OBJECTIVES OF THE STUDY



Research on pediatric patients with ALCAPA at Children's Hospital 1:

1. Determine the preoperative clinical features, ECG and transthoracic echocardiographic (TTE) presentations and anatomical characteristics perioperation.
2. Determine the characteristics of surgery and the rate of complications during surgery.
3. Determine the results of surgery through short-term and mid-term follow up: mortality rate, rate of complications (heart failure, pathological Q waves, LV function, mitral valve regurgitation, coronary arteries, supraaortic pulmonary stenosis)





# METHODS



- ❖ Retrospective and prospective case-series study
- ❖ All of ALCAPA patients was operated at Children's Hospital 1 from June 2008 to June 2020.



# SUBJECTS AND METHODS



**All patients with ALCAPA undergoing surgery met the following criteria:**

- ✓ ALCAPA patients are diagnosed by echocardiography, CCTA or ICA before treatment and the diagnosis were confirmed during surgery.
- ✓ There was a follow-up examination, echocardiography and electrocardiogram 1 month after surgery.
- ✓ Parents/guardians agreed to participate.



# SUBJECTS AND METHODS

## DEFINITION OF VARIABLES

- **In-hospital:** post-operative period in the hospital after the patient is treated with surgery.
- **Short-term period:** from discharge post-operation to < 24 months after surgery (re-examination at 1 month, 3 months, 6 months, 12 months, 18 ± 3 months).
- **Mid-term period:** from 24 months to 11 years after surgery (re-examination every year, from 2 years to 11 years).
- **Pathologic Q wave** was defined by duration  $\geq 0.03s$ , depth  $\geq 3 \text{ mm/V1}$  or  $\geq 2 \text{ mm/aVL}$

# RESEARCH FLOWCHART



## RETROSPECTIVE 6/2008 → 8/2014

- ICD10: Q24.5
- CICU, CATH LAB → ALCAPA operated

## PROSPECTIVE 8/2014 → 6/2020

- Cardiology department/Neonatal department/ ICU/NICU
- Diagnosed ALCAPA

## OPERATED ALCAPA Meets inclusion criteria

## DATA COLLECTION

- Clinical features and cardiac imaging results (ECG, Chest XRay, TTE, CCTA, ICA)
- Anatomical characteristics ALCAPA pre-operative
- Characteristics of surgery and the rate of complications during surgery.

## FOLLOW-UP POSTOPERATION

- Clinical examination evaluates heart failure
- Imaging: ECG, TTE, chest X-ray, CCTA/CMR/ICA when indicated.

**SHORT-TERM FOLLOW UP (< 24 MONTHS) MID-TERM FOLLOW UP (≥ 24 MONTHS)**  
Heart failure, pathological Q waves, LV function, mitral valve regurgitation, coronary arteries, supralvalvular pulmonary stenosis

Objective  
1 & 2

Objective  
3



# RESULTS - DISCUSSION



## ALCAPA: 51 CASES

- ✓ 10 cases: retrospective & prospective
- ✓ 41 cases: prospective
- ✓ Median follow-up duration: **36 (24 - 60)** months, maximum **108** months

**ALCAPA**  
**51 CASES**

### SHORT-TERM FOLLOW UP

1 month: N = 49  
< 24 months: N = 41

**In-hospital death: 2 cases**  
**Late death: 1 case**

### MID-TERM FOLLOW UP

24 months: N = 41  
>= 5 yrs: N = 19



# RESULTS - DISCUSSION



## PREOPERATIVE FEATURES

	Naimo Phillip, 2015	Shivani, 2017	Hui-Li, 2017	Our study
<b>N</b>	42	37	50	51
<b>Female</b>	-	-	21 (42%)	27 (52,9%)
<b>Age at operation (month)</b>	<b>4,7 (1,6 - 60)</b>	10.5 (1.3 – 219)	37,2 (4 – 216)	<b>4,5 (3,1 - 6,1)</b>
<b>Weight (Kg)</b>	5,7 (2,3 – 23)	-	<b>LVEF &lt; 50% group: 8,8 ± 3,1</b> LVEF > 50% group: 20 ± 13	5,2 (4,8 - 6)
<b>Infant type</b>	<b>34 (81%)</b>	<b>&lt; 1 year group: 54%</b>	<b>LVEF &lt; 50% group: 34 (68%)</b>	<b>44 (86,3%)</b>
<b>Severe HF (ROSS grade III &amp; IV)</b>	-	57%	-	<b>49 (94,1%)</b>
<b>Pathologic Q wave</b>			<b>80%</b>	<b>50 (98%)</b>



Johnsrude (1995), ECG → to distinguish Myocarditis and DCM from ALCAPA.  
 Zheng Jian-yong (2010): wide Q/DI **81,3%**, depth Q/aVL **100%** infant group < 12 ms.



## RESULTS - DISCUSSION

### ECHOCARDIOGRAPHIC FINDINGS



Echocardiographic markers	All patients (N = 51)	Infant type (N = 44)	Adult type (N = 7)	p
1. Visualization of coronary artery origin	46 (90,2%)	41 (93,2%)	5 (71,4%)	0,13
2. Abnormal Doppler signal in PA	45 (88,2%)	40 (90,9%)	5 (71,4%)	0,18
3. Flow reversal in LMCA	38 (74,5%)	35 (79,5%)	3 (42,9%)	0,06
4. Collateral coronary flow	48 (94,1%)	41 (93,2%)	7 (100%)	0,47
5. Z score RCA	5,41 ± 1,88	<b>5,20 ± 1,51</b>	<b>6,73 ± 3,25</b>	<b>0,044</b>
6. MR	<b>51 (100%)</b>	<b>44 (100%)</b>	<b>7 (100%)</b>	0,22
▪ Moderate MR	44 (86,3%)	38 (86,4%)	6 (85,7%)	
▪ Severe MR	5 (9,8%)	5 (11,4%)	0 (0%)	
7. LV systolic function	44 (86,3%)	44 (100%)	0 (0%)	
LVEF (%)	39,22 ± 14,20	35,02 ± 9,99	65,57 ± 4,96	
Z score LVEDD	5,53 ± 1,31	<b>5,75 ± 1,18</b>	<b>4,2 ± 1,31</b>	<b>0,002</b>
8. LV endocardial and papillary muscle fibroelastosis	<b>51 (100)</b>	<b>44 (100)</b>	<b>7 (100)</b>	

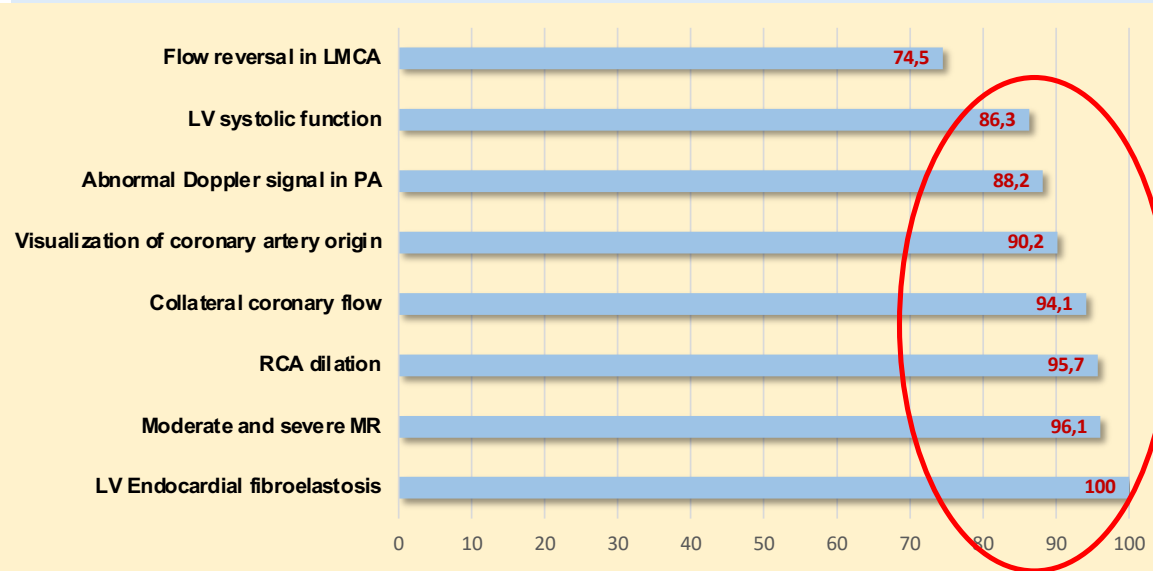
# RESULTS - DISCUSSION



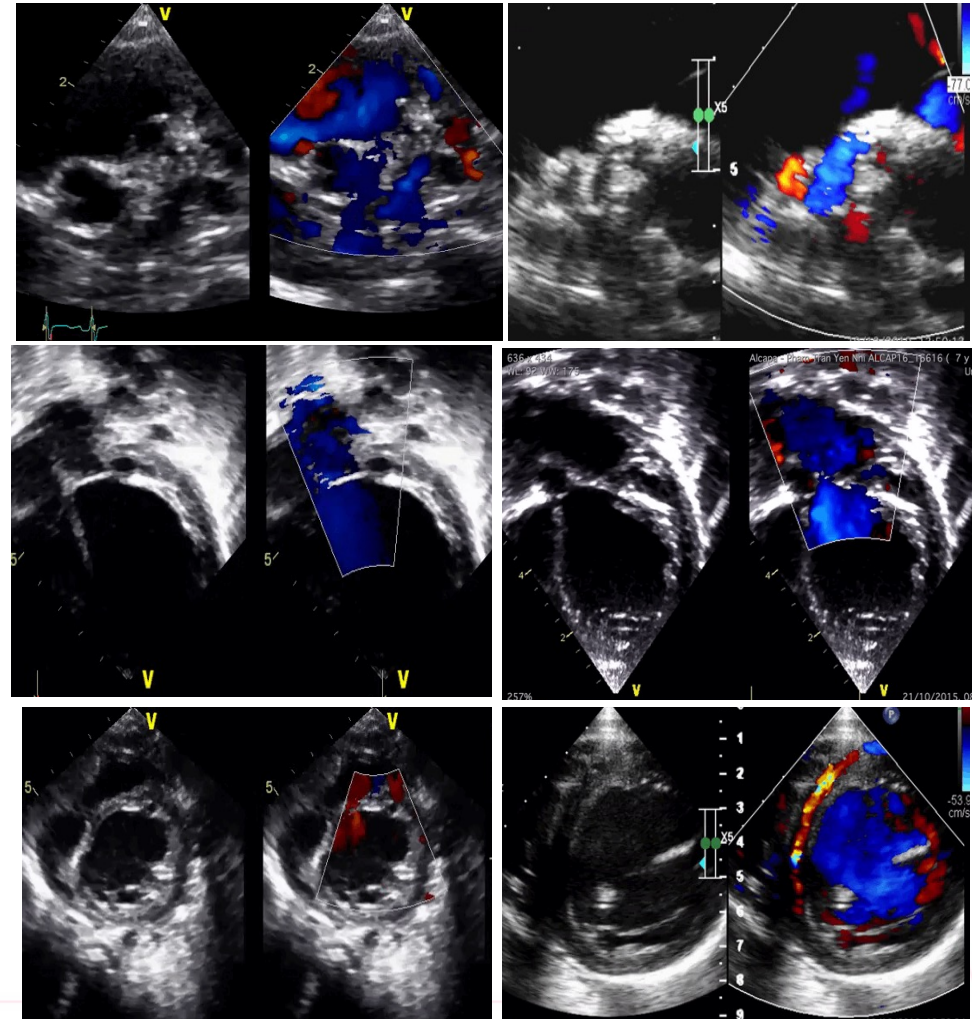
Thầy thuốc tận tâm - Chăm sóc tận tình

## PREOPERATIVE ECHOCARDIOGRAPHY

### INCIDENCE OF ECHOCARDIOGRAPHIC MARKERS OF ALCAPA



Shivani GP (2017), *J Am Soc Echocardiogr*, 30(9): N = 37, presenting echocardiograms had **five of seven** markers in **85%** of patients.  
Rong-Juan Li (2016), *Medicine (Baltimore)*, 95(15): **5/8** makers in **77,3%**

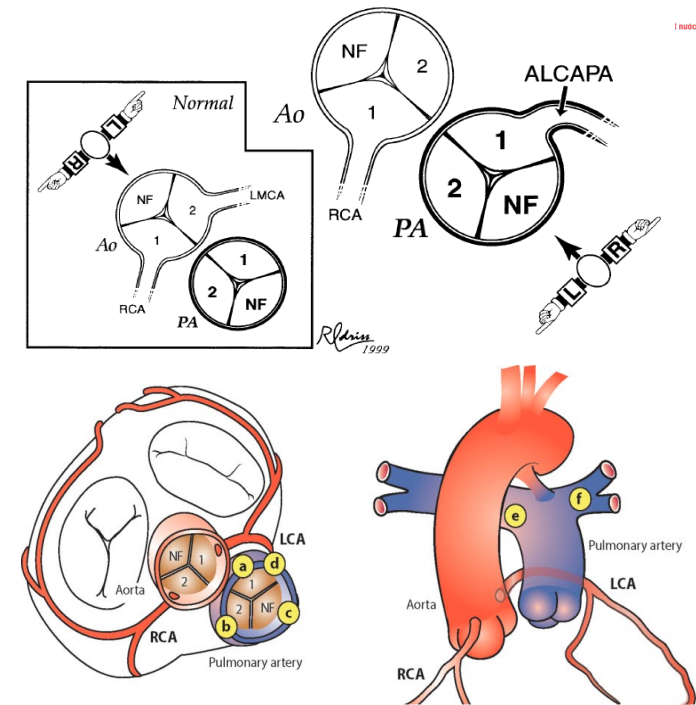


# RESULTS - DISCUSSION

## ANATOMICAL CHARACTERISTIC ALCAPA

Location of origin of the ALCAPA	TTE (N = 51)	CCTA (N = 26)	ICA (N = 10)	Operation (N = 51)
Right facing sinus of Valsalva, (n,%)	<b>42 (82,4)</b>	19 (73,1)	8 (80)	<b>38 (74,5)</b>
Nonfacing sinus of Valsalva, (n,%)	2 (3,9)	0 (0)	0 (0)	3 (5,9)
Posterior intercommissural region, (n,%)	5 (9,8)	1 (3,8)		4 (7,8)
MPA (n,%)	2 (3,9)	6 (23,1)	2 (20)	<b>6 (10)</b>

Kudumula Vikram (2014), *Ann Thorac Surg*, 97(3): N1 sinus of PV: **15 (60%)**  
 Weigand Justin (2015), *Pediatr Cardiol*, 36(3), pp. 489-97: N1 sinus of PV: **23 (52%)**



Karimi M. (2015), *World J Pediatr Congenita Heart Surg*, 6(4) 526-540

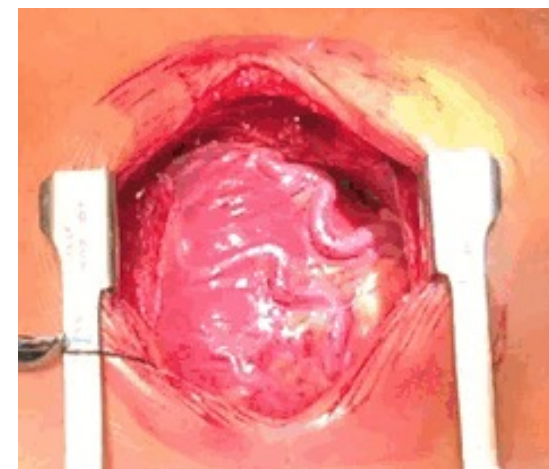
**TTE:** Transthoracic echocardiography  
**CCTA:** Coronary computed tomography angiography  
**ICA:** Invasive coronary angiography

# RESULTS - DISCUSSION

# CHARACTERISTICS OF ALCAPA SURGERY



SURGICAL OPTIONS	Result (N = 51)
<i>Re-establish dual-coronary artery system</i>	51 (100%)
▪ Coronary button transfer	<b>38 (74,5%)</b>
▪ Coronary button transfer with PA flap	<b>13 (25,5%)</b>
MV repair in initial operation	<b>3 (5,9%)</b>
Ventricular fibrillation durring surgery	<b>20 (39,2%)</b>
Cross-clamp time (min)	61,5 ± 28,1
CPB time (min)	126,5 ± 26,5



## ❖ Coronary button transfer procedure

- Azakie (Toronto, 2003): 47/67 (70%)
- Turley (1995), Barth: Coronary button transfer and tubular reconstruction, reduce anastomotic tension, reduce coronary artery stenosis and PA stenosis.

## ❖ MV repair

- Isomatsu (Japan): 3,7%, annuloplasty)/severe MR

# RESULTS - DISCUSSION

# POSTOPERATIVE/CICU - MORTALITY



Thầy thuốc tận tâm - Chăm sóc tận tình

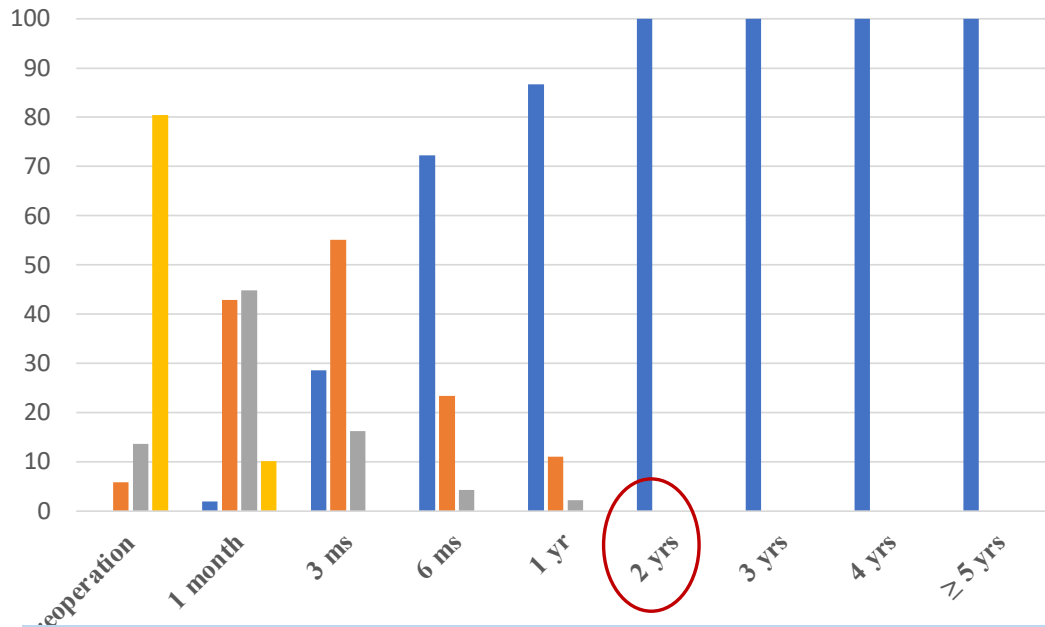
Postoperative characteristics	Result (N = 51)
Postoperative ICU stay (day), median (range)	9 (7 - 13)
Postoperative ventilation time (h), median (range)	<b>120 (76 - 168)</b>
Prolonged mechanical ventilation $\geq 5$ ds, n (%)	<b>44 (86,3%)</b>
<b>Low cardiac output syndrome (n,%)</b>	<b>19 (37,3%)</b>
Postoperative ventricular arrhythmia	11 (21,6%)
Duration of inotropic support (h), median (range)	<b>156 (112 – 196)</b>
Prolonged duration of inotropic support $\geq 6$ ds, n (%)	<b>31 (60,8%)</b>
<b>Mortality</b>	
▪ Early mortality (n,%)	<b>2 (3,9%)</b>
▪ Late mortality (n,%)	<b>1 (1,9%)</b>

- ❖ **Postoperative ICU stay:**
  - Hui-Li (China, 2017): 96 (45 - 720) hours
  - Ojala Tiina (Finland, 2010): 8 ds
- ❖ **Mechanical ventilation time:**
  - Hui-Li (China, 2017): **49,5** (10 - 504) hours
- ❖ **Low cardiac output syndrome (LCOS)**
  - Samuel (Mexico, 2011): **38%**
- ❖ **Early mortality:**
  - Naimo Philip (Australia, 2015): 1/42 (**2,4%**)
  - Yan Gao (China, 2017): 2/26 (7,6%)
- ❖ **Late mortality:**
  - Yan Gao (China, 2017): 1/26 (3,8%)
  - Antonio (2015): 1/34 (**3%**)

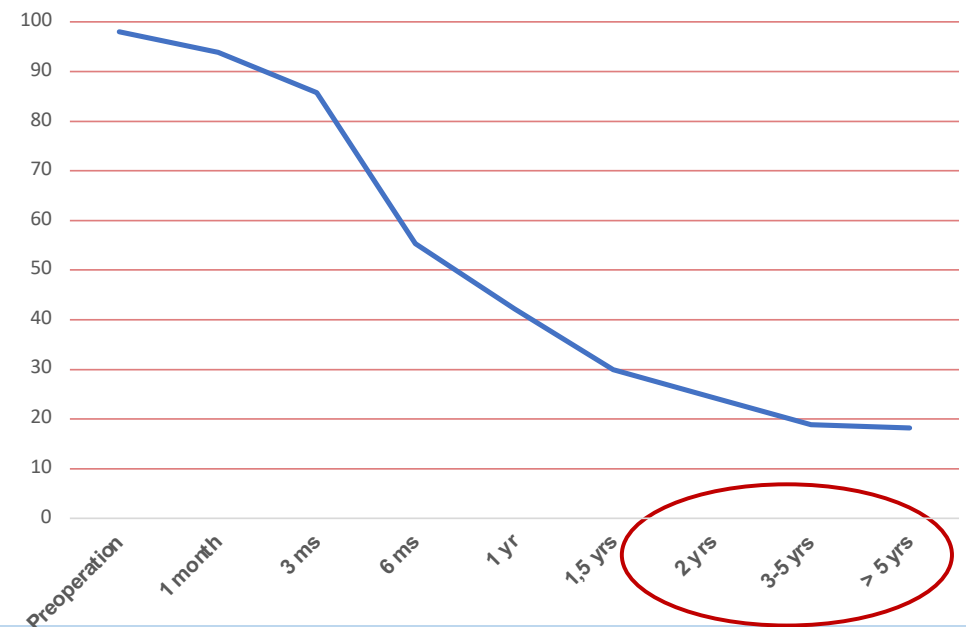
# RESULTS - DISCUSSION



**PROGRESSION OF HEART FAILURE AFTER ALCAPA SURGERY**



**PROGRESSION OF PATHOLOGIC Q WAVE AFTER ALCAPA SURGERY**



- ❖ **Pathologic Q wave** recovers slowly, after > 5 years: 18.2%, slower than other studies even though we follow-up longer
- ❖ Zheng Jian-yong (2010), *Chin Med J (Engl)*, 123(20): pathologic Q wave disappeared by mean follow up time **17 ms (6 - 166)**
- ❖ Hsin-Hui (2008), *Eur J Pediatr*, 167(11): 13 patients, pathologic Q waves disappeared after median of **20 months (1 - 48)**

# RESULTS - DISCUSSION

## LEFT VENTRICLE FUNCTION



Thầy thuốc tận tâm - Chăm sóc tận nước

	Antonio, 2015	Muna, 2020	Our study
N	34	29	51
Follow up time (month)	72 (1 – 168)	60 ± 43,2	36 (24 – 60)
<b>Z score LVEDD</b>			
Preoperation	6 ± 3,8	5,7 ± 2,8	<b>5,53 ± 1,31</b>
Last examination	0,9 ± 0,7	0,1 ± 0,7	<b>0,11 ± 0,65</b>
<b>LVFS (%)</b>			
Preoperation	25 ± 14	15 ± 10	<b>18,86 ± 8,18</b>
Last examination	38 ± 5	37 ± 3,1	<b>36 ± 2,8</b>
<b>LVEF (%)</b>			
Preoperation	21 ± 6	34 ± 17	<b>39,22 ± 14,2</b>
Last examination	60 ± 7	63 ± 4,1	<b>67,1 ± 3,7</b>

**LVEF:** Left ventricular ejection fraction  
**LVEDD:** Left ventricle end-diastolic dimension  
**LVFS:** Left ventricular fractional shortening

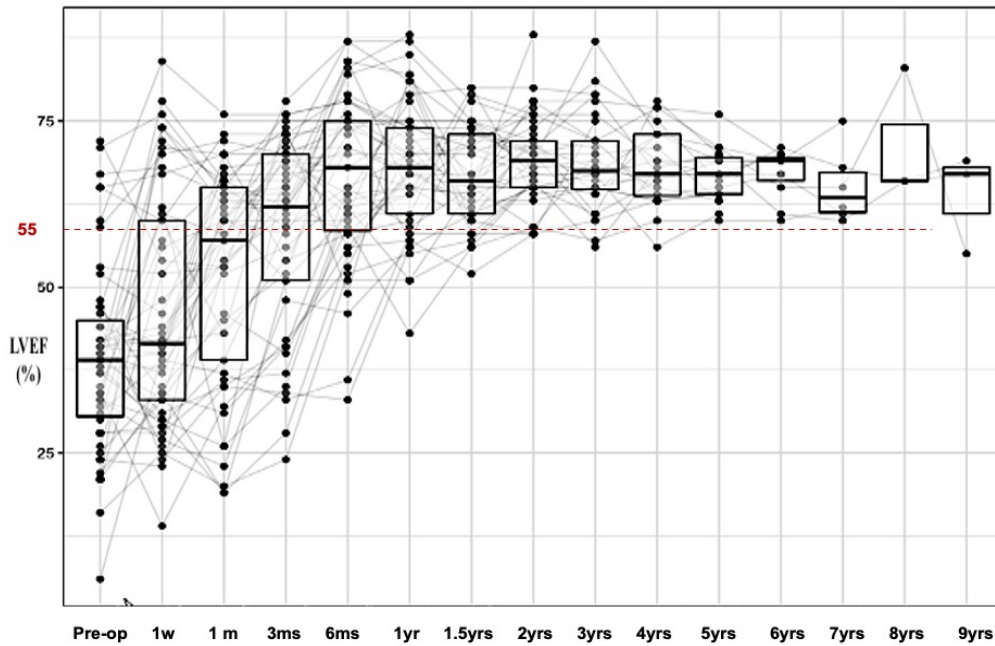


# RESULTS - DISCUSSION

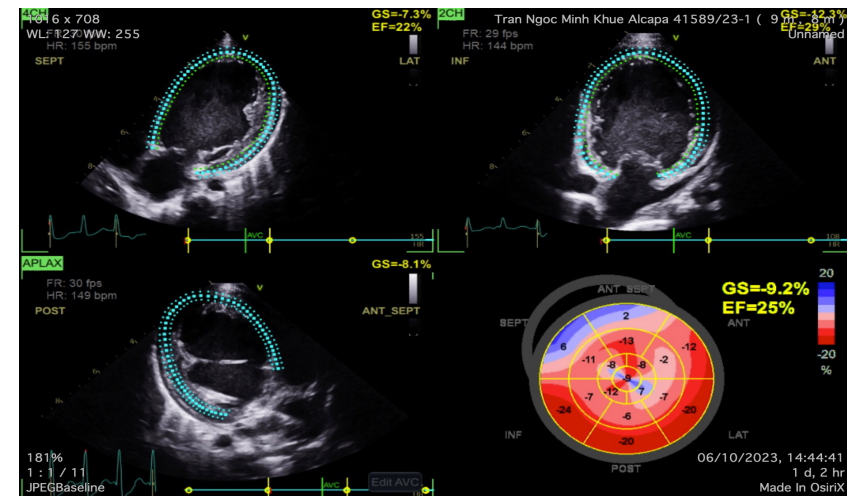
## LEFT VENTRICLE FUNCTION



### Progression of LVEF after ALCAPA surgery



LVEF recovered quickly in the first 6 months after surgery and the curve stabilized to normal values after 6 months.



LVEF: Left ventricular ejection fraction



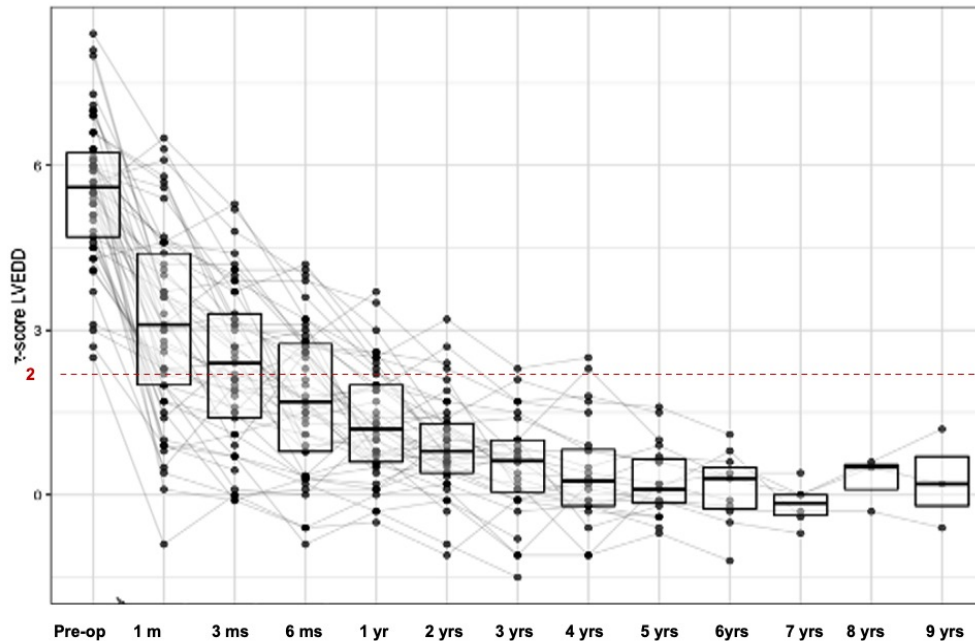


# RESULTS - DISCUSSION

## LEFT VENTRICLE FUNCTION



### Progression of LVEDD Z score after ALCAPA surgery

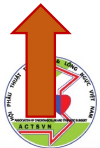


LVEDD Z score decreased rapidly in the first 6 months after surgery, and LVEDD reached to almost normal value 1 year after surgery.

The LVEDD Z score recovered more slowly than LVEF because the LV were dilated due to mitral valve regurgitation.

Mid-term follow-up showed that LVEDD Z score recovered to normal value, approximately 0 value about 4 - 5 years after surgery.

LVEDD: Left ventricle end-diastolic dimension



# RESULTS - DISCUSSION



## Recovery time of LVEF in infant type group

	Justin, 2015	Yan Gao, 2017	Muna, 2020	Our study
<b>N</b>	44	26	29	44
<b>Follow up time (month)</b>	32 (17 – 114)	40 (3 – 96)	60 ± 43,2	36 (24 – 60)
<b>LVEF recovery time (month)</b>	3,03 (0,17 – 14,3)	5,3 (0,5 – 36)	1,5 – 12	<b>3,0 (1,0 – 6,1)</b>

- **Infant type group (N = 44)**

- ✓ ≥ 6 months: **92,9%** LVEF normalized

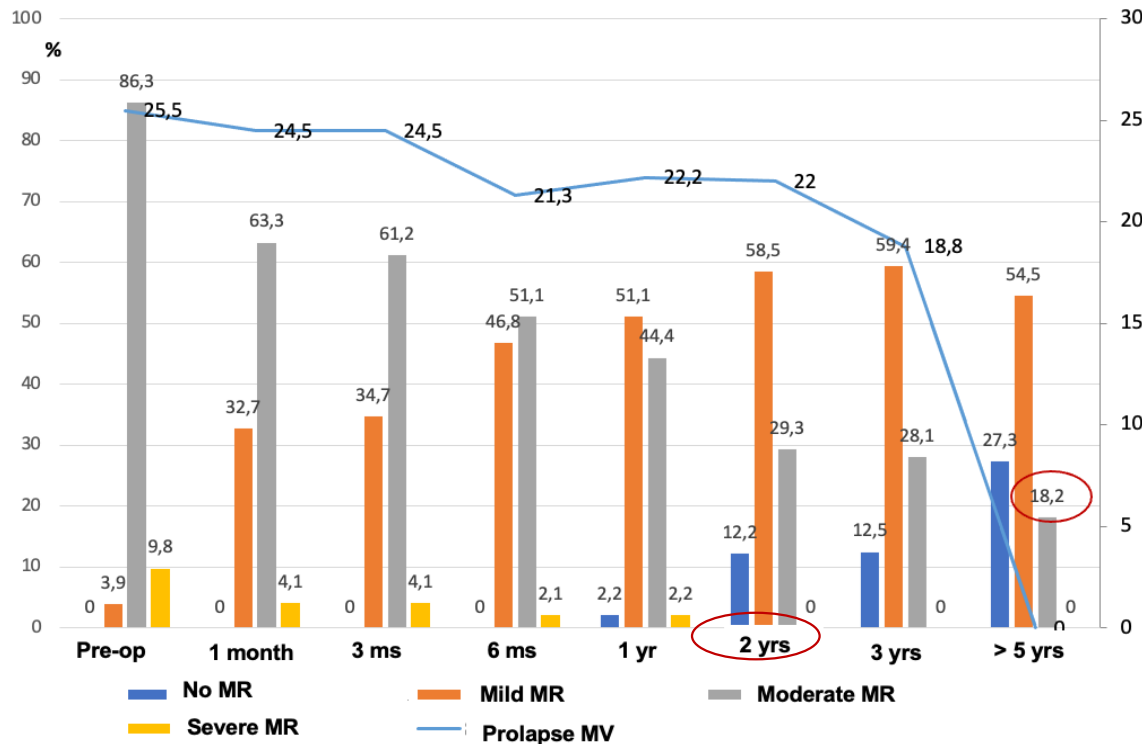
- ✓ 12 months: **100%** LVEF recovered as normal value

- Deepa Sasikumar (2018), *Interact Cardiovasc Thorac Surg*, 27(2): **3 months, 88%** LV function normalized

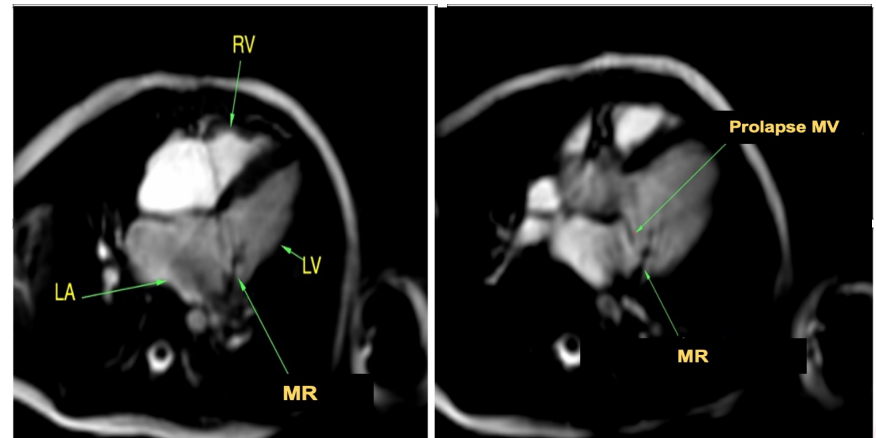
# RESULTS - DISCUSSION

## MITRAL REGURGITATION

PROGRESSION OF MR AFTER ALCAPA SUGERY



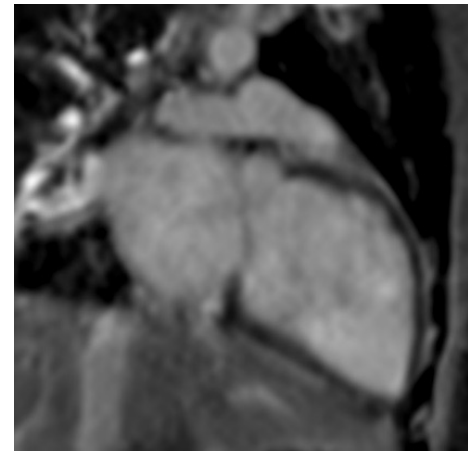
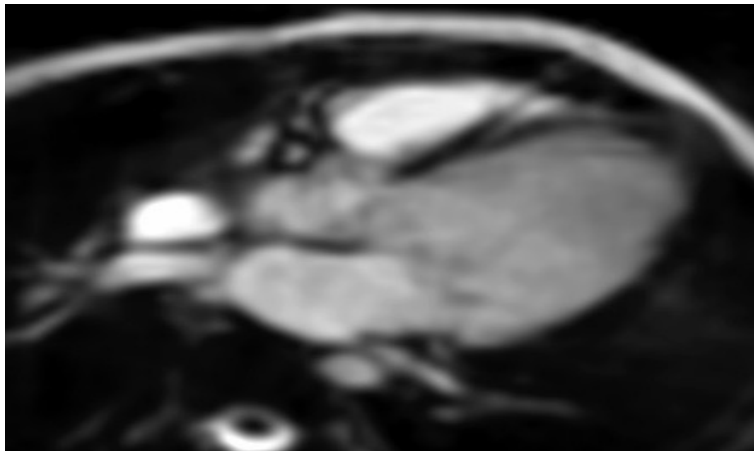
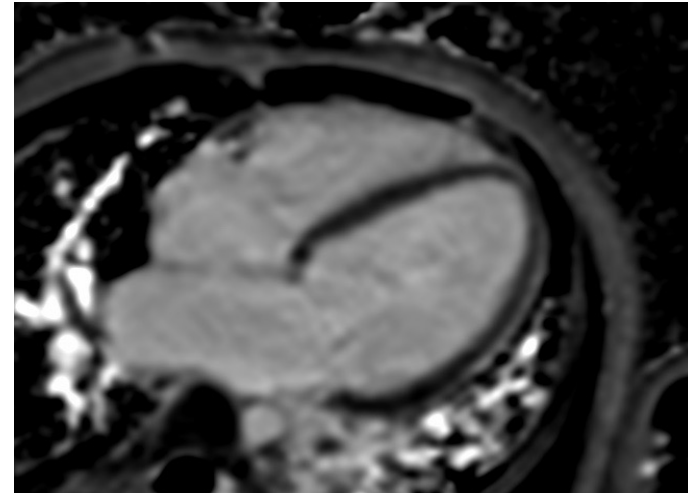
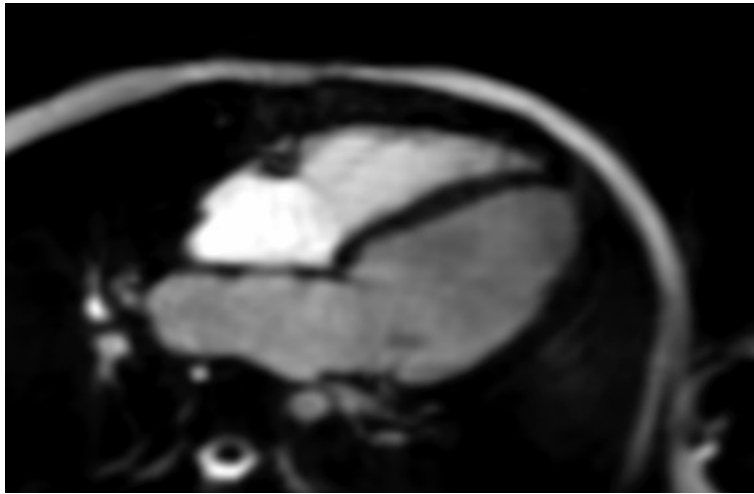
- LV endocardial and papillary muscle fibroelastosis
- ✓ 2 years: **100%** patients
- ✓ 5 – 9 years: **81,8%** patients
- Vladimir Alexi (2011): **57%**, LV myocardial scarring/CMR



# RESULTS: MR/CMR



Thầy thuốc tận tâm - Chăm sóc tận tình



# RESULTS - DISCUSSION



## MITRAL REGURGITATION

	John Brown, 2008	Vikram Kudumula, 2014	Antonio Cabrera, 2015	Our study
N	25	25	34	51
Follow up time (months)	84 (1 – 192)	93 (9 – 240)	72 (1 – 168)	36 (24 – 60)
<b>MR (n,%)</b>				
No MR		8/21 (38,1%)	12/17 (71%)	4/32 (12,5%)
Mild MR	22 (88%)	9/21 (42,9%)	5/17 (29%)	19/32 (59,4%)
Moderate MR	2 (8%)	4/21 (19%)	9 (26%)	9/32 (28,1%)
Severe MR	1 (4%)	0	2 (6%)	0%
<b>MV repair</b>				
Initial operation	<b>0%</b>	<b>4 (16%)</b>	<b>5 (15%)</b>	<b>3 (5,9%)</b>
Re-operation	<b>3 (12%)</b>		<b>1 (3%)</b>	<b>0%</b>

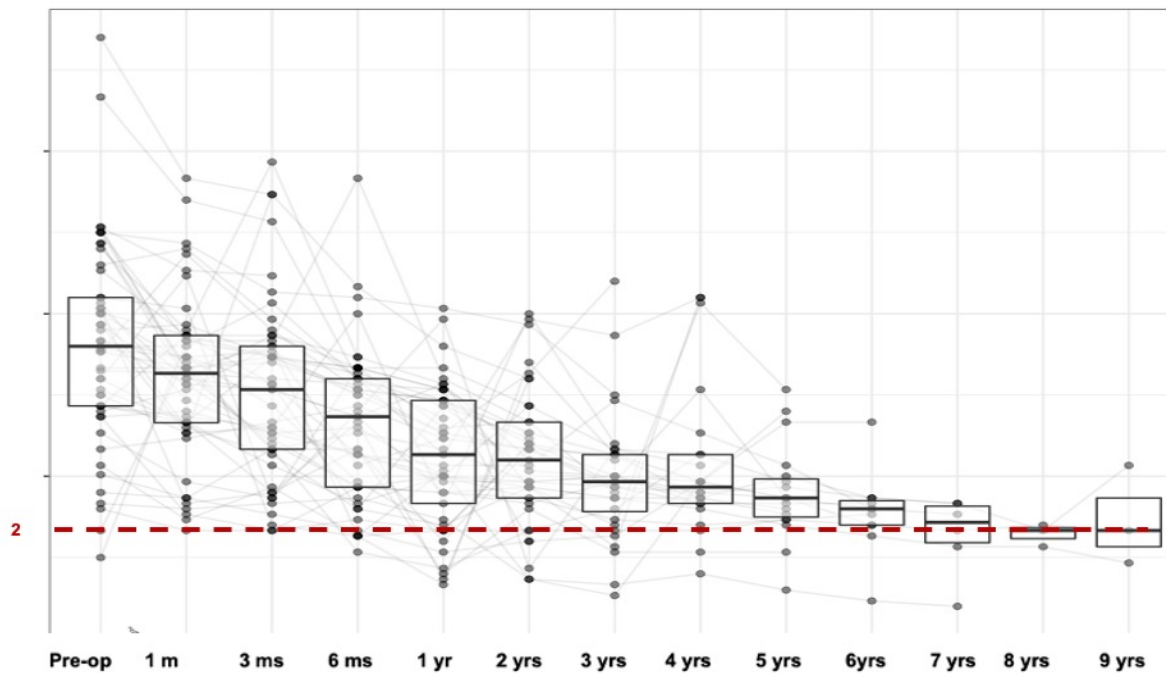
- Deepa Sasikumar (2018), *Interact Cardiovasc Thorac Surg*, 27(2): the rate of MV repair in older group (25%) **higher than** < 1 year group (5,9%)

# RESULTS - DISCUSSION



## CORONARY ARTERY

### PROGRESSION OF Z SCORE RCA POSTOPERATIVE



#### ❖ *Left coronary artery:*

- ✓ No LCA stenosis
- ✓ No fistula/baffle leak

#### ❖ *RCA compensatory dilation*

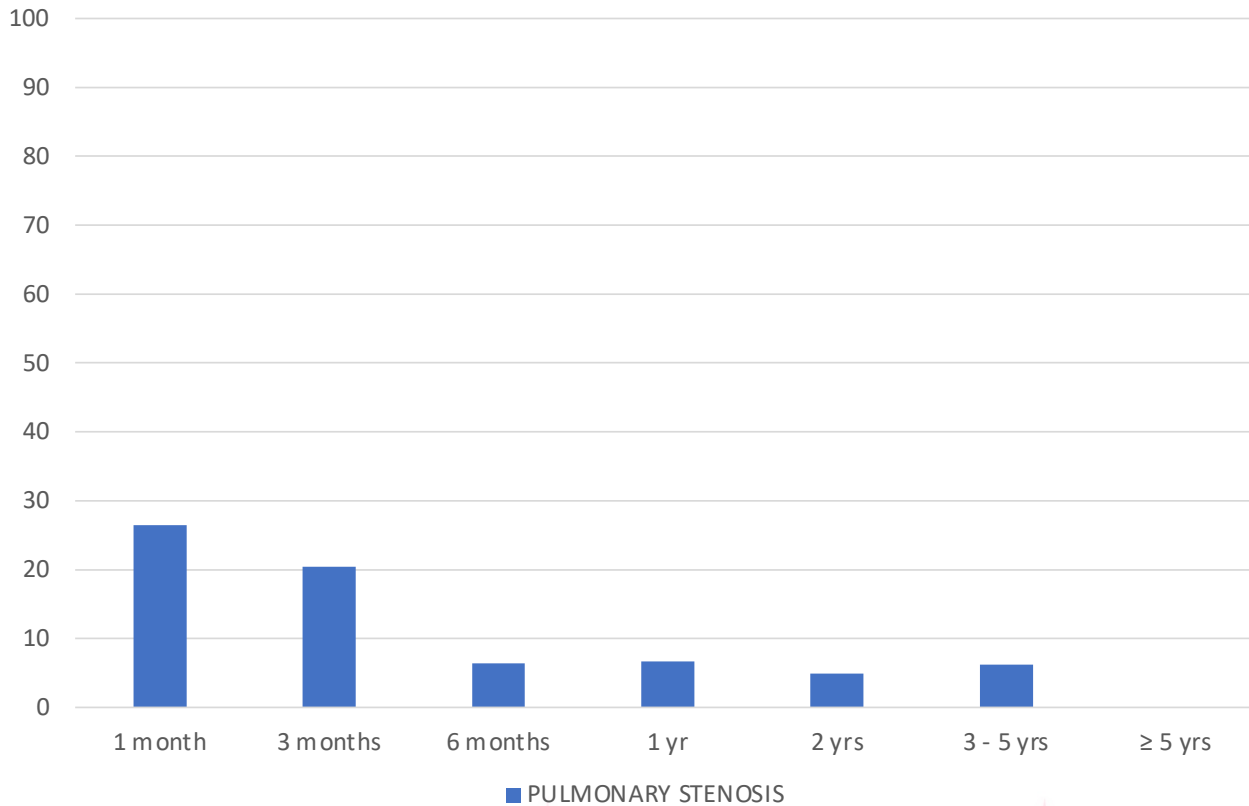
- Gradually reduce size after surgery
- Did not return to normal size



# RESULTS - DISCUSSION



## PULMONARY STENOSIS



### ❖ *The rate of PS:*

- Guido Michielon (2003), *Ann Thorac Surg*, 76(2): 3/31 **(9,7%)**
- Yan Gao (2017), *Chin Med J (Engl)*, 130(19) : 6/26 **(26,1%)**



# CONCLUSIONS



The study of **51 ALCAPA patients** underwent surgery at Children's Hospital 1 with a median follow-up time of **36 (24 - 60) months**, maximum **108 months**.

- ✓ The median age at surgery: 4,5 (IQR: 3.1-6.1) months, median weight: 5.2 (IQR: 4.8-6) Kg.
- ✓ Infant types were the majority and the rate of severe heart failure before surgery is high
- ✓ Combination echocardiographic makers of ALCAPA and ECG were valuable in diagnosis
- ✓ MV annuloplasty: 3 (5.9%) at initial ALCAPA repair.
- ✓ The rate of early complications were high
  - Ventricular fibrillation 39.2%,
  - Low cardiac output syndrome 37.3%





# CONCLUSIONS



## ➤ Short-term follow up

- ✓ Mortality rate 5,9%
- ✓ All children had no symptoms of heart failure and restored LV function on echocardiography
- ✓ LVEF normalized at a median of 3.0 (IQR: 1.0 – 6.1) months.

## ➤ Mid-term follow up

- ✓ All children were free of pulmonary valve stenosis
- ✓ 18,2% of patients had pathologic Q waves and moderate MR persisted for more than 5 years post-operation
- ✓ Most of the patients (81.8%) remained with enhanced echogenicity of papillary muscles and LV endocardium
- ✓ The RCA dilation was prolonged for more than 5 years post-operation



**THANK YOU FOR YOUR ATTENSIION!**

