学校法人　東京女子医科大学
Tokyo women's Medical University

Surgical repair for Tetralogy of Fallot

Tokyo Women's Medical University
Department of Cardiovascular Surgery

Takeshi Shinkawa

4th Asian Association for Pediatric and Congenital Heart Surgery; June 1st, 2024

Tetralogy of Fallot

- First cyanotic lesion described
- First palliative and definitive operations

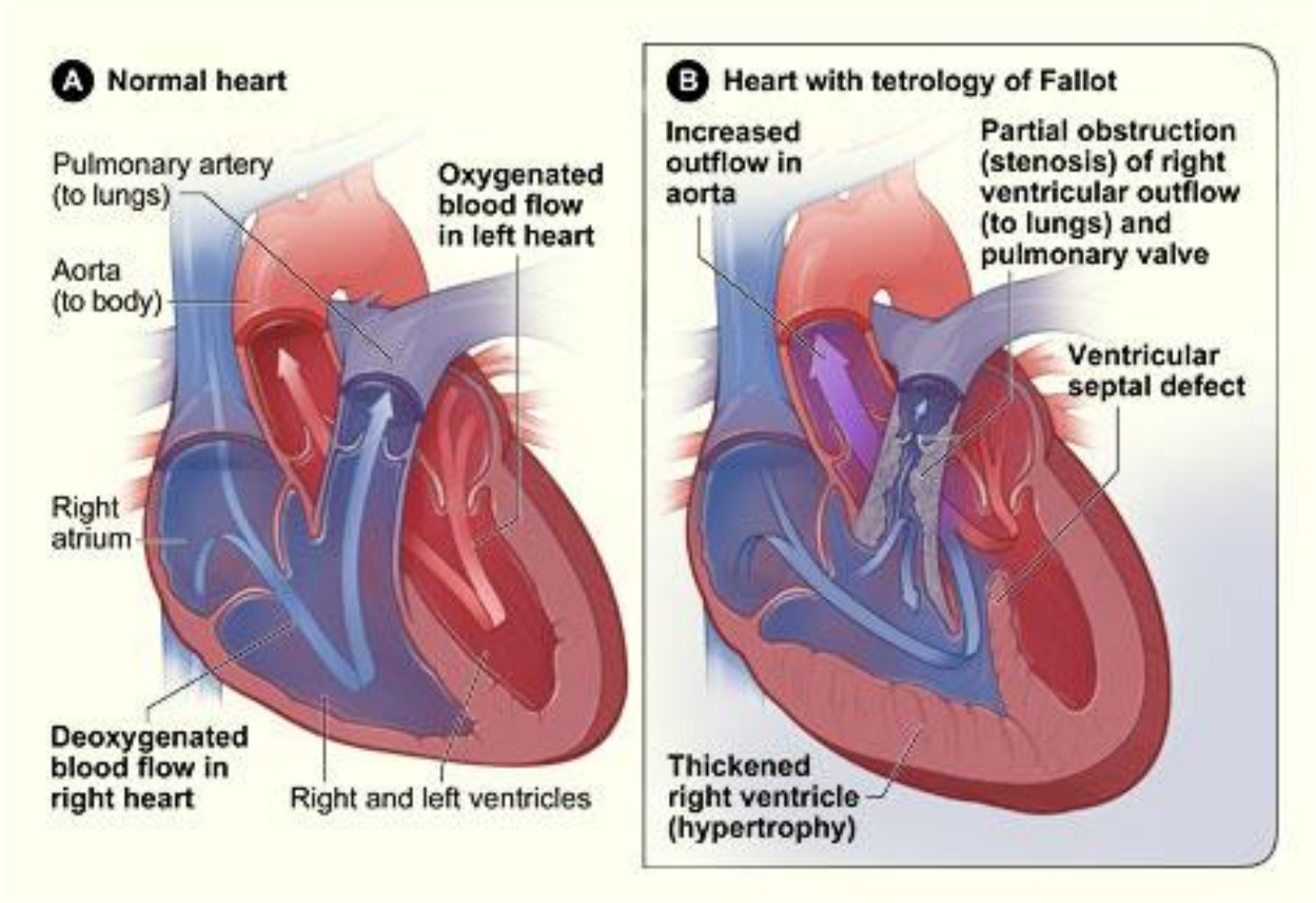
- First cohort of long term survivors
- Model for natural history studies of treated CHD
- Central role in molecular biology and myocardial protection

- Lethal if untreated, good surgical results, but still a challenge
- Requiring a life-long management

Anatomy

- VSD
- RVOT obstruction
- Aortic override

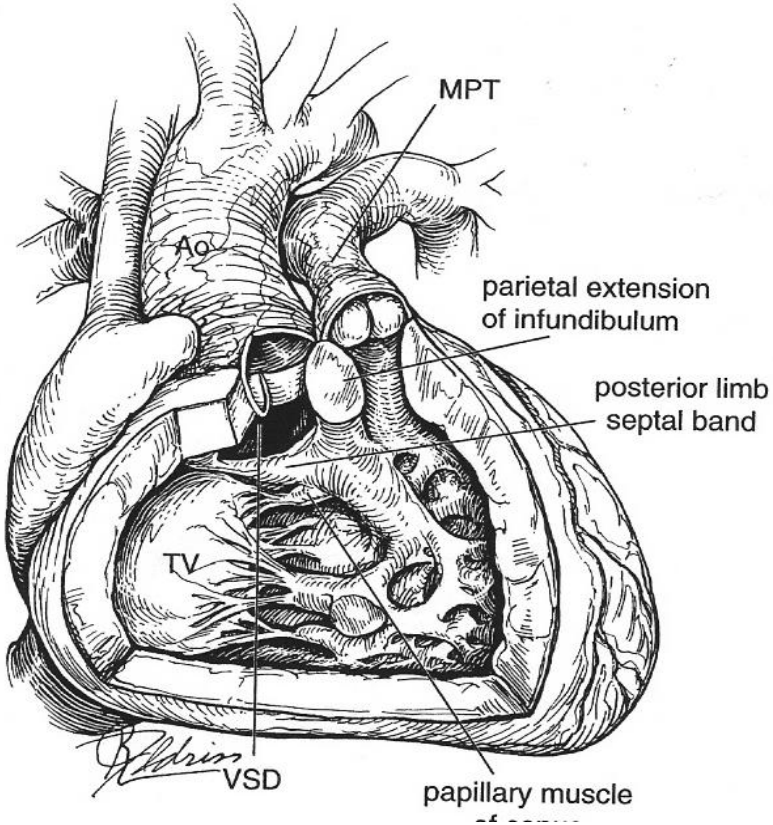
- RV hypertrophy



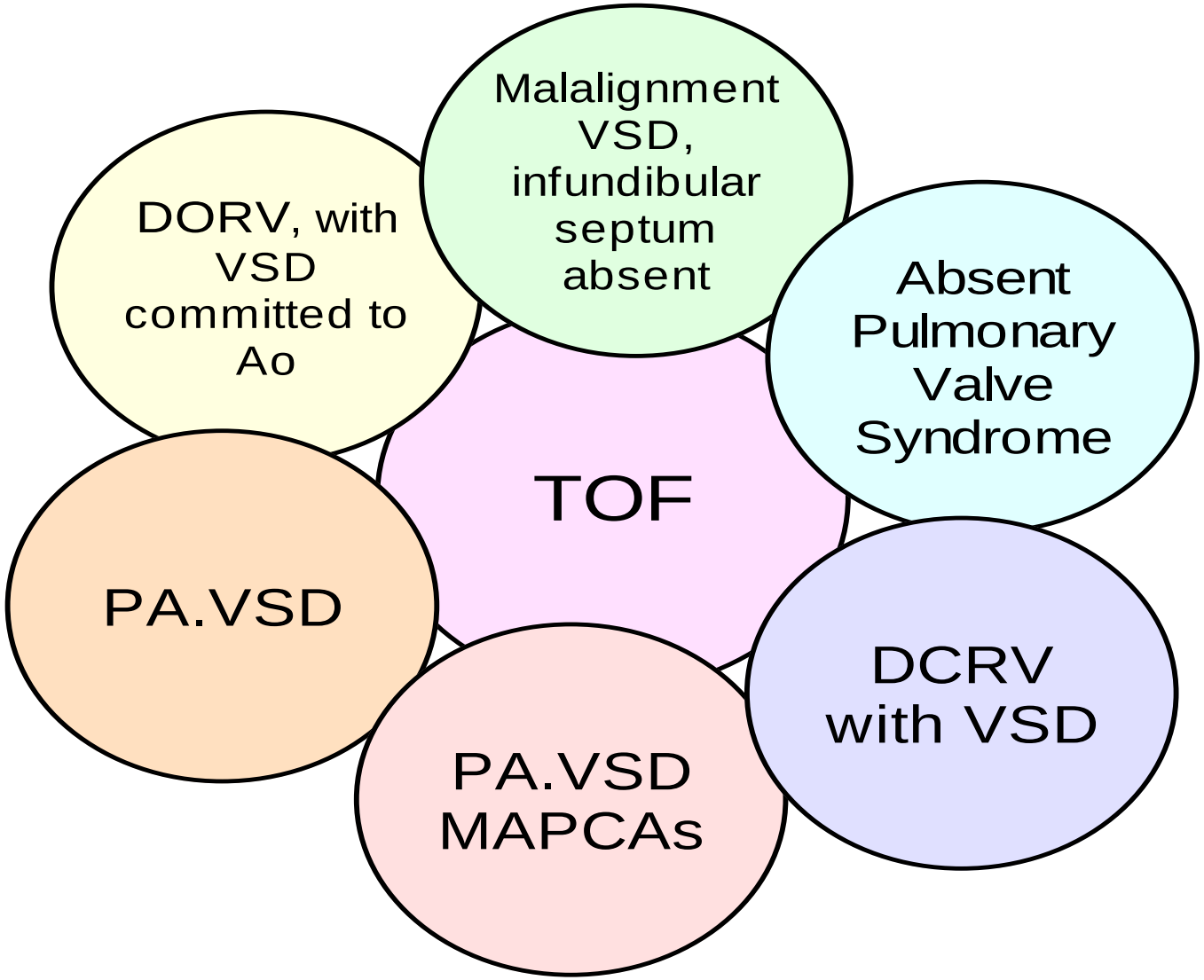
Anatomy

- Aortic valve moves to the left and anterior: Overrides
- RVOT obstruction from infundibular “crowding”
- VSD creation
- RV hypertrophy is acquired

- Pulmonary valve abnormalities
  - Smaller annulus
  - Thick leaflets, tethered to PA wall
  - Bicuspid valve in up to 60% of pts
- PA hypoplasia

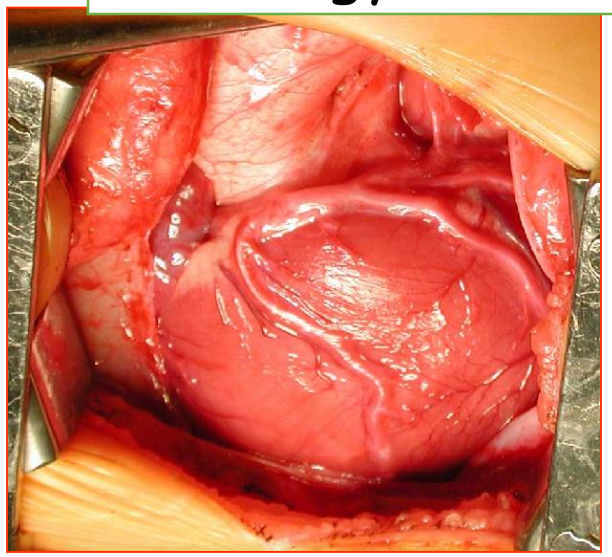


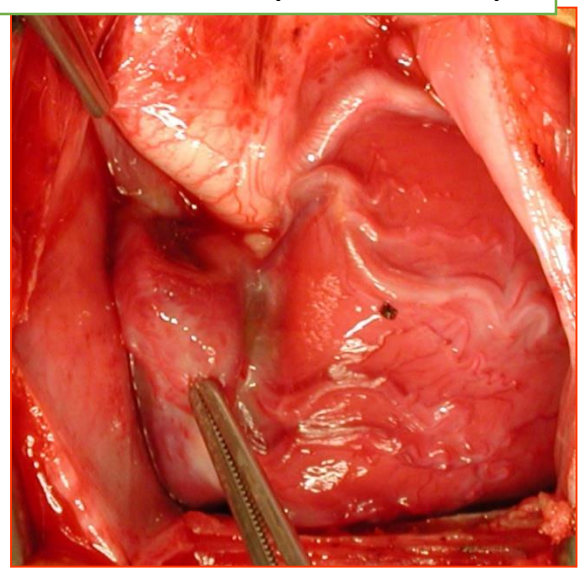
Anatomy


Malalignment VSD, infundibular septum absent
DORV, with VSD committed to Ao
Absent Pulmonary Valve Syndrome
TOF
PA.VSD
DCRV with VSD
PA.VSD MAPCAs

Anatomy

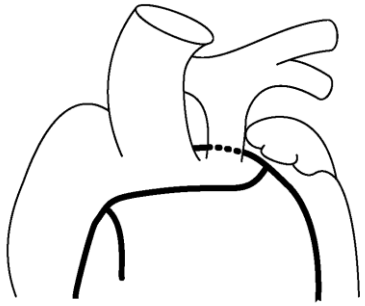
Tetralogy with abnormal coronaries (36/611)



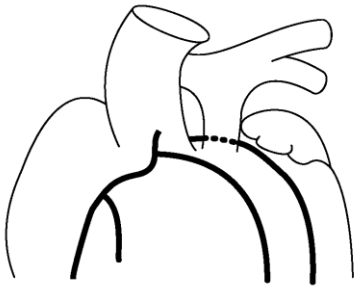


RAD (22)

R from LAD (8)



Dual AD-Conal (6)



History

- 1671
  - N Stensen: First description in malformed fetus
- 1671-1888
  - Numerous case presentation (Sandifort, Hunter, etc)
- 1888
  - E Fallot: Non-random association of the tetrad
    leading to specific symptomatology
- 1924
  - M Abbott: Tetralogy of Fallot

History

- 1944
  - A Blalock: Palliation – shunt
- 1954
  - CW Lillehei: Intracardiac repair with cross-circulation
- 1955
  - JW Kirklin: Intracardiac repair with pump oxygenator

The Blalock-Taussig Shunt

© Karsh

One of the photographic studies done by Yousuf Karsh, to mark the official "1,000th blue baby" procedure performed by Dr. Alfred Blalock.

© Karsh

Vivien Theodore Thomas: surgical technician

The Blalock-Taussig Shunt

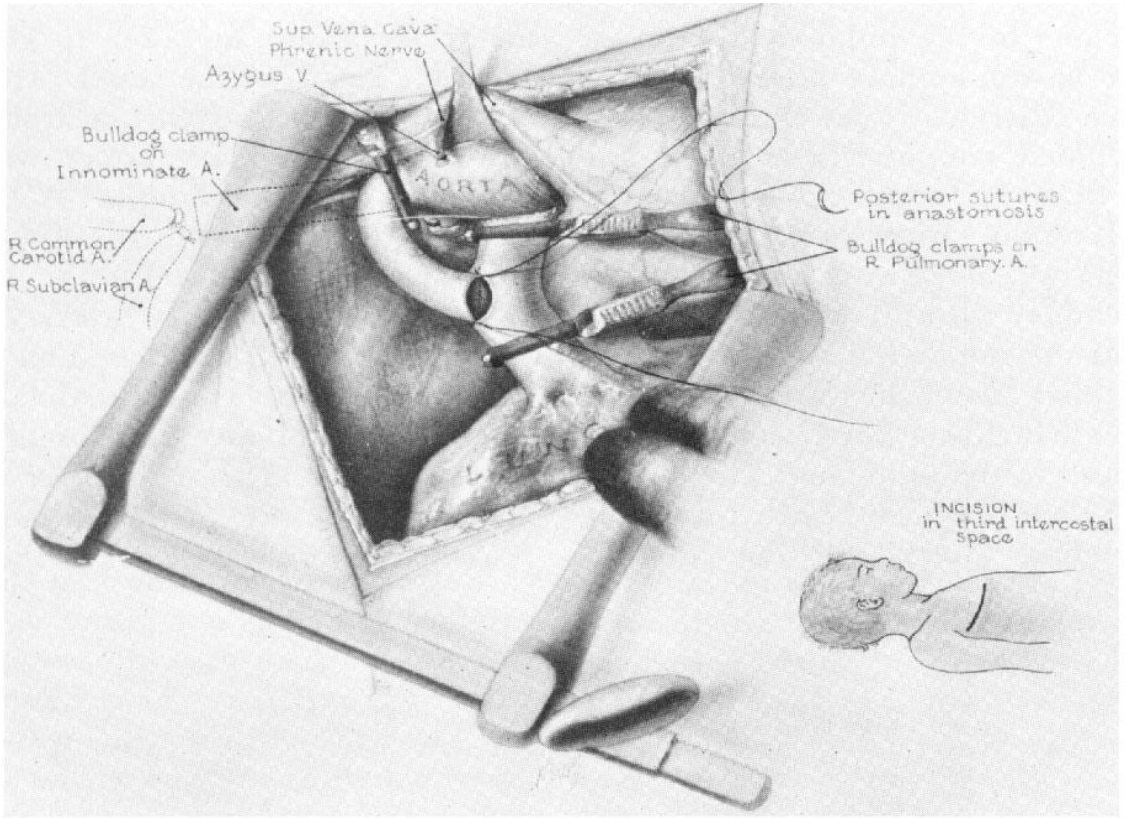


FIG. 1. General exposure of the operative field on the right side. The end of the innominate artery is being anastomosed to the side of the right pulmonary artery. The posterior row of sutures is complete. The anterior row has not been inserted.

HLH 4 E Surgery

E S

OPERATION: Nov. 29, 1944
Dr. Alfred Blalock
Ether - Oxygen - Dr. Harmel

ANASTOMOSIS OF LEFT PULMONARY ARTERY TO LEFT SUBCLAVIAN ARTERY

This patient was an undernourished child who had cyanosis on frequent occasions. The diagnosis was pulmonary stenosis.

Under ether and oxygen, administered by the open method, an incision was made in the left chest extending from the edge of the sternum to the axillary line in the third interspace. The second and third costal cartilages were divided. The pleural cavity was entered. The left lung looked normal. No thrill was felt in palpating the heart and pulmonary artery. The left pulmonary artery was identified and was dissected free of the neighboring tissues. The left pulmonary artery seemed to be of normal size. The superior pulmonary vein, on the other hand, seemed considerably smaller than normal to me. I had hoped that the artery to the left upper lobe might be sufficiently long to allow an anastomosis, but this did not appear to be the case. The left subclavian artery was then identified and was dissected free of the neighboring tissues. The vertebral artery and the branches of the thyrocervical axis were doubly ligated and divided. The subclavian was so short that there would not have been sufficient length for our purposes, had this not been done. The subclavian artery was then ligated distal to the thyrocervical trunk. A bulldog clip was placed on the subclavian artery at a point just distal to its origin from the aorta. The subclavian artery was then divided just proximal to the ligature. Two bulldog clips were then placed on the left pulmonary artery, the first clip being placed at the origin of the left pulmonary artery, and the second clip being placed just proximal to the point where the artery entered the lung. There was ample space between these two clips for our purpose. A small transverse incision was then made in the wall of the pulmonary artery. By the use of chine beaded silk on fine needles, an anastomosis was then performed between the end of the left subclavian artery and the side of the left pulmonary artery. A posterior row of sutures was placed first. There was practically no bleeding following the removal of the bulldog clips.

The anastomosis seemed to be a satisfactory one, and the main point of worry comes from the small size of the left subclavian artery. I was disturbed because I could not feel a thrill in the pulmonary artery after the clips were removed. I do not believe that this was due to any clot in the subclavian artery, because it seemed to pulsate vigorously. It is possible that it was due to a low pressure in the systemic circulation. I do not actually know what the systemic pressure was. Another possibility was that it might have been due to spasm of the subclavian artery. My only regret was that the subclavian artery was not bigger. It is possible that the increased red cell count in this patient may have predisposed to thrombosis.

(over)

Sulfanilamide was placed in the left pleural cavity. This was followed by closure of the incision in the chest wall. The third and fourth ribs were approximated by two encircling sutures of braided silk. The soft tissues of the wall were closed in layers with silk sutures.

The patient stood the procedure better than I had anticipated. It is interesting that the cyanosis did not appear to increase very greatly from the temporary occlusion of the left pulmonary artery. It is also of interest that the circulation in the nail beds of the left hand appeared to be fairly good at the completion of the operation.

I did not attempt to visualize the left common carotid artery. It is possible that this would have been bigger than the left subclavian. This child was very small and I am confident that the subclavian artery would be more easily dealt with in a larger child.

(Dr. Blalock)

ms

Op Report
Nov 29, 1944

Assistants: Dr W. Longmire
Dr D. Cooley

History

- 1944
  - A Blalock: Palliation – shunt
- 1954
  - CW Lillehei: Intracardiac repair with cross-circulation
- 1955
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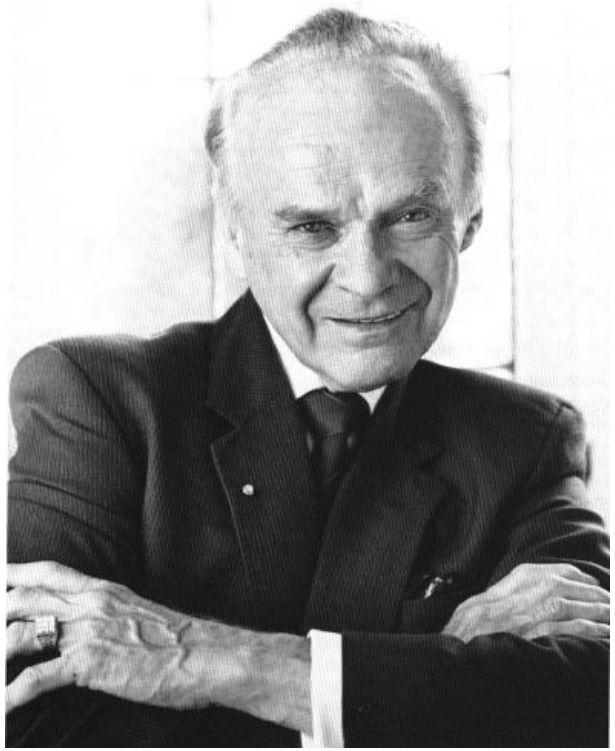
Initial TOF repair



The First Open Heart Corrections of Tetralogy of Fallot

A 26–31 Year Follow-up of 106 Patients

C. WALTON LILLEHEI, PH.D., M.D.
RICHARD L. VARCO, PH.D., M.D.
MORLEY COHEN, PH.D., M.D.
HERBERT E. WARDEN, M.D.
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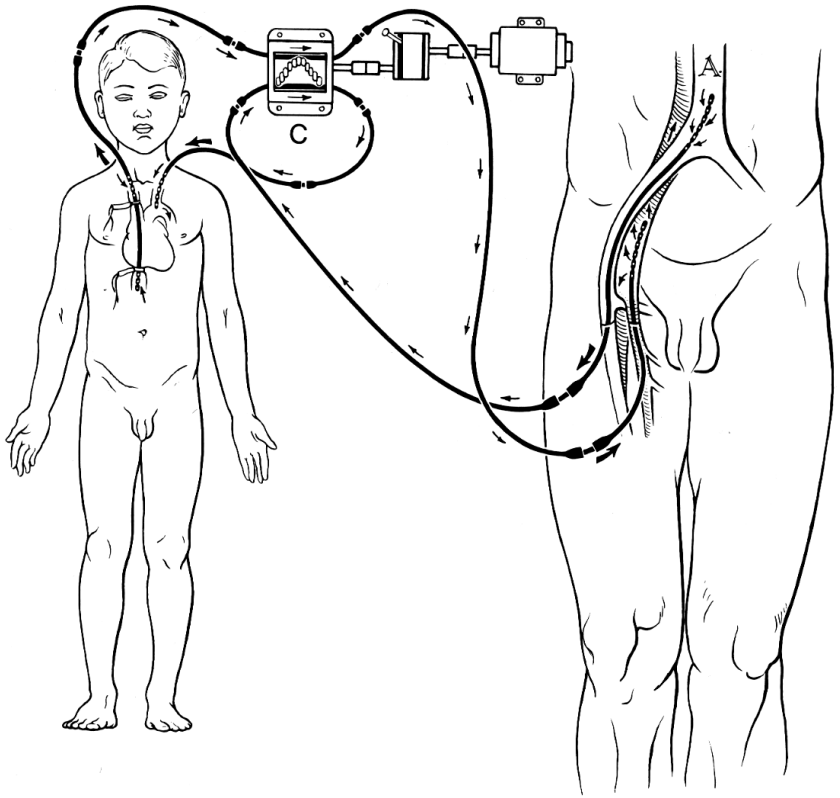
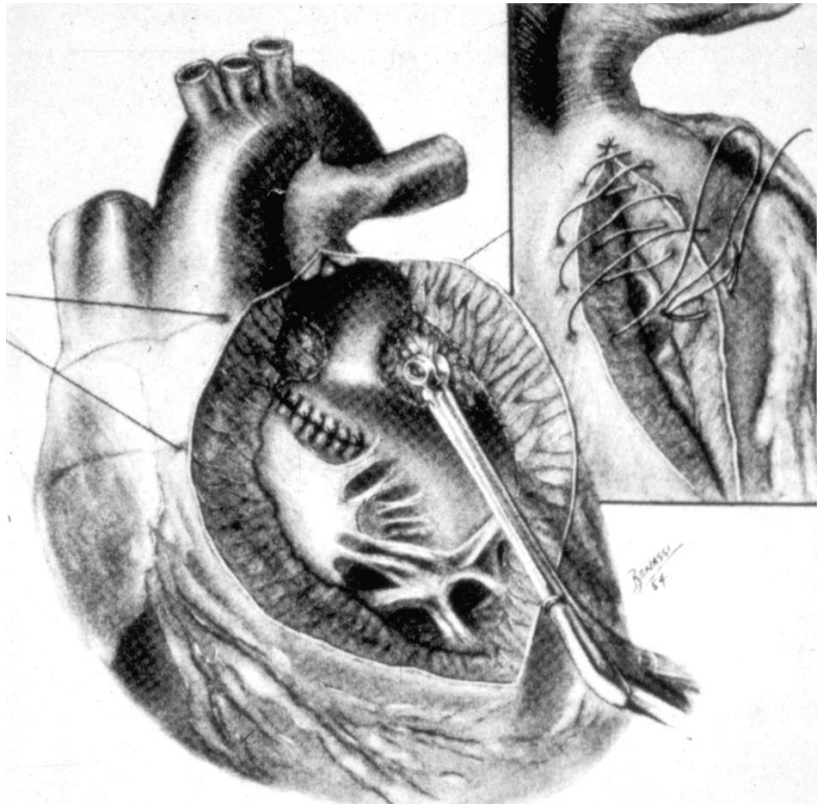




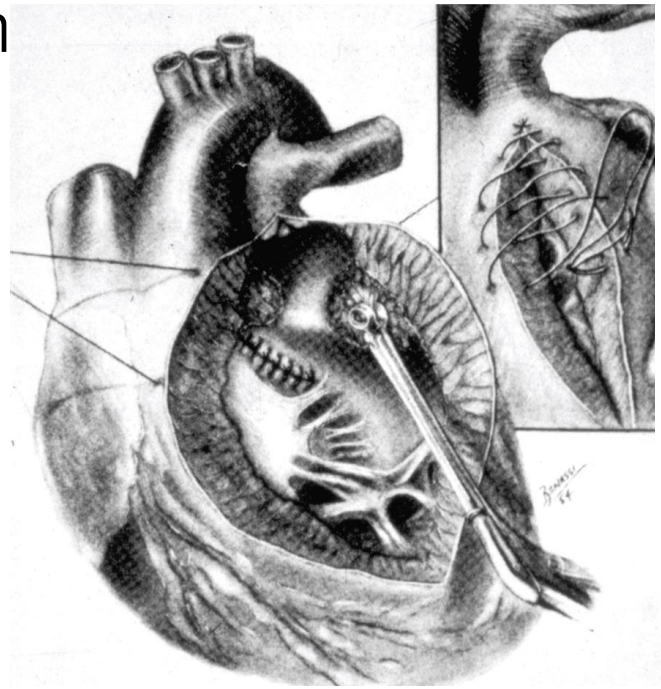
Ann Surg. 1986;204:490-502.

Initial TOF repair

- 106 pts (4 months-45 years)

- Survival 77% at 30 yrs
- Freedom from re-operation
  91% at 30 years

- 32% college
- 10% graduate school



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From the University of Minnesota Hospitals and Variety C Heart Hospital, Minneapolis, Minn

Tetralogy of Fallot became a correctable malformation on August 31, 1954, and from that date through 1960, 106 patients (ages 4 months–45 years) who underwent open repairs at the University of Minnesota and were discharged, have been followed (99% complete) until death or for 26–31 years (mean: 23.7 years, 2424 patient years). The purposes of this study were to determine survival, morbidity, hemodynamics, educational/employment attainments, and relation of these to surgical technics. Operations were done by cross circulation (6 patients) and bubble oxygenator (100 patients). This group had the first uses of patch ventricular septal defect closure, outflow root, infundibuloplasty, atresia correction, ischemic arrests, and pacemakers among other innovations. Twenty-one (of 105 patients) have died during the followup: eight deaths in the first 10 years, 12 between 10 and 20 years, and 1 >20 years. The causes of death were sudden (5), accidental (4), congestive failure (2), reoperation (2), suicide (2), and other (2). Actuarial survival at 30 years was 77%. Late complications were ten reoperations, five arrhythmias, and one endocarditis. Actuarial freedom from reoperations at 30 years was 91%. Cardiac recatheterizations in 62 patients disclosed only 10 with residual shunts. Peak right ventricular systolic pressures were <40 mmHg (34 patients), 41–60 mm (2 patients), 61–70 mm (4 patients), >71 mm (4 patients). Thirty-four patients (32%) completed college, ten of these completed graduate school (5 masters degrees, 2 M.D.'s, 2 Ph.D.'s, 1 lawyer). Fifteen others attended college, and nine received technical school diplomas. Forty patients (18 men, 22 women) had progeny, with 82 (93%) live births and six major cardiac defects (7.3%). In summary, complete repair gave excellent late results in this group cared for very early in the open heart era. Survivors led productive lives without restrictions in education and employment. Many of the deaths/complications that occurred are now easily preventable, which augurs extremely well for this generation.

Second thoughts are ever wiser.
from Euripides, 4th Century, B.C.

Presented at the 106th Annual Meeting of American Surgical Association, Hot Springs, Virginia, April 24–26, 1986.

Reprint requests: Dr. C. Walton Lillehei, 73 Otis Lane, St. Paul, MN 55104.

Submitted for publication: July 3, 1986.

TETRALOGY OF FALLOT (TOF), a common form cyanotic heart disease in infants and childr became a correctable malformation on Aug 31, 1954, when open cardiotomy with extracorporeal culation by controlled cross-circulation was utilized. first operation by these methods was successful. and patient is living an active, normal life as a professio musician 31.5 years after operation.[1]

The corrective procedures consisted of closing the tricular septal defect and relieving the pulmonary steno The intracardiac operation was, at that time, a signific departure from the palliation afforded by systemic-p monary artery shunts and thus provoked considera discussion at the time of our first report of a series of patients in April 1955.[2] As noted in that presentation, were sufficiently impressed by the well being of those o patients to make the intracardiac corrective operation method of choice, with very few exceptions, for all T patients referred for surgical treatment from that time

The purposes of this study were to determine surv morbidity, hemodynamics, child bearing, educational employment attainments, and the relation of these to gical techniques. Operations were done by controlled c circulation[3–5] (6 patients) and DeWall–Lillehei bub oxygenator[6–8] (100 patients). This patient group had first uses of patch ventricular septal defect (VSD) clos outflow roof, infundibuloplasty, atresia correction, i emic arrest, and pacemakers among other innovatio

Patients and Methods

The patients reported herein were all of those who intracardiac repair of their TOF defects from August

Ann Surg. 1986;204:490-502.

Conotruncal repair

- VSD closure via RV tomy
- Trans-annular patch
  with momocusp

Conotruncal Repair of Tetralogy of Fallot

Hiromi Kurosawa, M.D., Yasuharu Imai, M.D., Makoto Nakazawa, M.D.,
Kazuo Momma, M.D., and Atsuyoshi Takao, M.D.

ABSTRACT **A repair within the conotruncal portion of the right ventricle is introduced. It was used for the intracardiac repair of 30 consecutive patients with tetralogy of Fallot. The infundibular septum was totally resected to reduce the muscular outflow stenosis. Short patch infundibuloplasty with a large monocusp was then used. Instead of the tricuspid septal leaflet, the membranous flap was employed as the suture line for patching the ventricular septal defect (VSD) to avoid a conduction disturbance, residual VSD, and fixing of the tricuspid septal leaflet. The right ventricular (RV) to systemic arterial pressure ratio was 50.0 ± 14.6% (N = 26) and right atrial pressure was 9.0 ± 2.5 mm Hg (N = 26) one month after operation. RV end-diastolic volume was 93.0 ± 30.5% of normal (N = 15) before operation and 96.7 ± 29.0% of normal one month after operation in the same patients. These data suggest that a conotruncal repair can maintain good RV function with low right atrial pressure and with no increase of RV volume.**

Approximately ten years ago, we developed a standardized patch infundibuloplasty procedure for tetralogy of Fallot in which a patch infundibuloplasty 30 to 45% of the length of the right ventricle and a minimum infundibulectomy are sufficient to relieve the right ventricular (RV) outflow narrowing, while still preserving RV function [1]. The ventricular septal defect (VSD) is usually guarded on its inferior margin by the extension of the trabecula septomarginalis, and has a membranous flap that can be safely used for suture placement [2]. To achieve goals of low right atrial pressure, no heart murmur, and no conduction disturbance, we have endeavored to make the length of the patch infundibuloplasty as short as possible and to use a membranous flap for patching the VSD. The repair is performed entirely within the conotruncal portion of the right ventricle (Fig 1). Here we discuss this new method and its surgical results.

Material and Methods

From 1985 to 1986, 30 consecutive patients with common

pulmonary artery index [3] ranged from 159 to 360 cm^2/m^2 (average, 210 cm^2/m^2). Four patients had previously undergone palliative procedures including aorto-pulmonary shunt and RV outflow tract enlargement.

After the initiation of total bypass using extracorporeal circulation, a small, longitudinal incision was placed in the free wall of the RV outflow tract just beneath the pulmonary annulus. The incision was extended beyond the pulmonary annulus into the main pulmonary artery; the anterior commissure of the pulmonary valve was divided carefully in an effort to preserve valve function. The incision was extended distally onto the left pulmonary artery when necessary. Then the incision was extended caudally above the infundibular ostium of the right ventricle. This ventriculotomy was always just 30% of the length of the right ventricle; this length had been calculated prior to operation with our formula [1]. The ventriculotomy was retracted and kept open with four stay sutures (Fig 2A). Through this short ventriculotomy, only the tiny infundibular ostium and infundibular septum were seen. The inlet part of the right ventricle was not visualized.

Then the infundibular septum was totally resected. The incision began around the infundibular ostium from both sides of the infundibular septum at the junctions with the ventriculo-infundibular fold and with the anterior limb of the trabecula septomarginalis. The incision ran from the surface to the deep interior of the tissue, just short of the aortic valve. As a consequence, the infundibular septum was resected along the aortic sinus of Valsalva wall. During this procedure, the endocardium remained a few millimeters beneath the pulmonary valve and later was used as the suture line for VSD closure, since the aortic sinus of Valsalva wall had been partly denuded and was not strong enough to hold sutures (Fig 2B).

To avoid any damage to the aortic valve and wall of the sinus of Valsalva just behind the infundibular septum, good exposure was necessary and hence, a short ventriculotomy was mandatory. Without a short ventriculotomy, this particular resection procedure is difficult through either the tricuspid or pulmonary valve. A short ventriculotomy was also essential for enlargement of the

Ann Thorac Surg. 1988;45:661-6.

Conotruncal repair

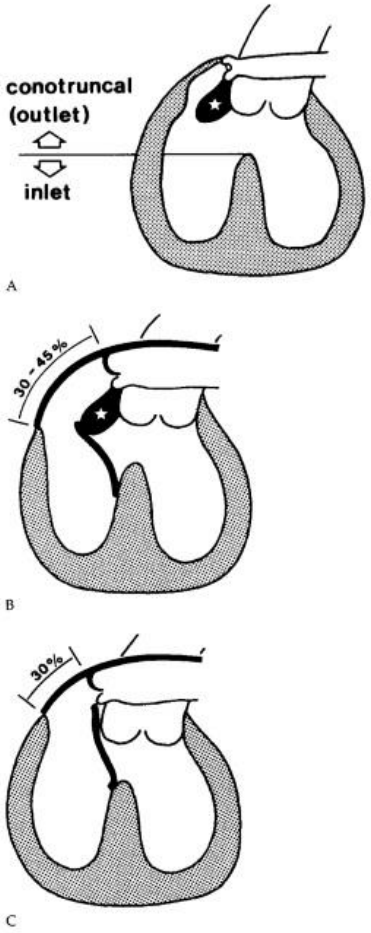
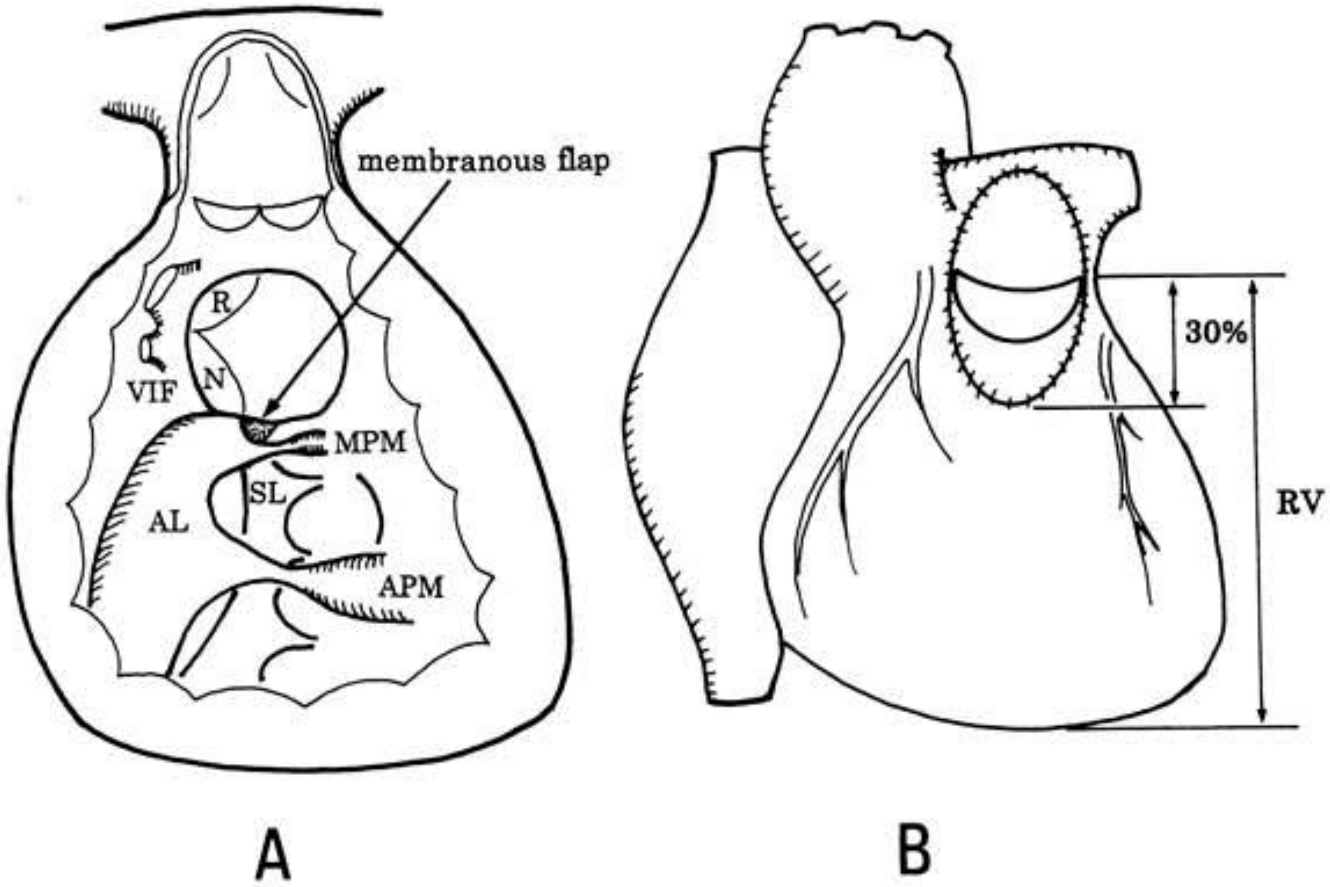


Fig 1. (A) In tetralogy of Fallot, right ventricular (RV) outflow narrowing is caused by the anterior displacement of the infundibular septum (white star) and the dextroposition of the aorta. (B) Stan-

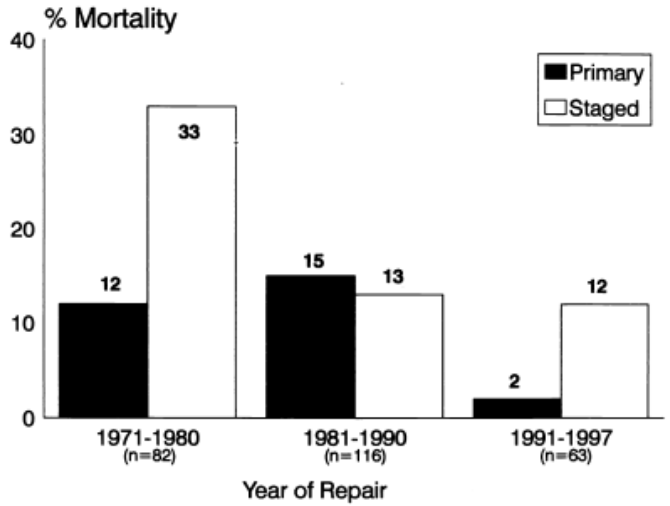
Ann Thorac Surg. 1988;45:661-6



J Thorac Cardiovasc Surg. 1998;115:351-60.

Early and late outcomes

- Knott-Craig et al 1998
  - 294 patients (1971-97)
  - 20 yr survival for hospital survivor
    : 0.98 (+/-.07)
  - freedom from re-intervention
    : 0.86 (+/- .04)



A 26-Year Experience With Surgical Management of Tetralogy of Fallot: Risk Analysis for Mortality or Late Reintervention

Christopher J. Knott-Craig, MD, Ronald C. Elkins, MD, Mary M. Lane, PhD, Jeannie Holz, RN, Carolyn McCue, RN, and Kent E. Ward, MD

Sections of Thoracic Surgery and Pediatric Cardiology, University of Oklahoma Health Sciences Center, Oklahoma City, Oklahoma

Background. Over the past decade repair of tetralogy of Fallot (TOF) in infancy has gained favor. It is still uncertain what effect early complete repair will have on survival or late reoperation on the right ventricular outflow tract.

Methods. To assess these outcomes, we reviewed our experience (1971–1997) with 294 patients undergoing operation at one institution. Median follow-up was 10.6 years (range, 0.1 to 26 years), and was complete for 90.2% patients.

Results. Primary complete repair was done in 199 patients (68%), and a staged repair in 62 patients (21%). Thirty-three patients had only a palliative procedure. Sixty-eight patients (23.1%) had complex pathologic processes, including pulmonary atresia in 53. Hospital mortality for primary repair was 11.1% (22/199), for staged repair was 17.7% (11/62), and for palliative procedures was 15.5% (16/103 procedures). Since 1990 mortality has been 2.1%, 11.8%, and 0% respectively ($p < 0.001$), despite younger age at repair (0.6 ± 0.1 versus 2.1 ± 0.2 years; $p < 0.001$). Multivariate analysis identified longer period of hypothermic circulatory arrest, pulmonary artery patch angioplasty, earlier year of operation, and closure of the foramen ovale as risk factors for hospital death. For hospital survivors 20-year survival was 98% ± 3% for TOF with pulmonary stenosis and 88% ± 9% for TOF with pulmonary atresia ($p = 0.09$). Reintervention on the right ventricular outflow tract was needed in 14.1% (37/261) patients. Freedom from reintervention on the right ventricular outflow tract at 20 years was 86% ± 4% for TOF with pulmonary stenosis and 43% ± 16% for TOF with pulmonary atresia ($p = 0.001$). For the subgroup TOF with pulmonary stenosis, this was 85% ± 5% after primary repair and 91% ± 8% after staged repair (not significant). At 15-year follow-up, this was 78% ± 10% for patients not older than 1 year at operation compared with 88% ± 4% for older patients (not significant).

Conclusions. Early mortality after primary repair of TOF has significantly improved and late survival is excellent. Primary repair in infancy does not increase risk for reintervention on the right ventricular outflow tract.

(Ann Thorac Surg 1998;66:506–11)

Tetralogy of Fallot (TOF) is the most common cyanotic congenital heart disease with approximately 3,000 new cases diagnosed in the United States each year [1]. Since the first successful repair by Lillehei and colleagues in 1954 [2], several reports have documented the excellent long-term prognosis for these patients, at the same time recognizing that those repaired after about 7 years of age fared less well [1, 3–6].

Despite some advocates of routine primary repair in infancy [7, 8], unacceptable early mortality resulted in wide acceptance of a two-stage repair, with excellent results [9, 10]. During the early 1990s, reports documented improved early results with primary repair in infancy [11–19] and suggested that primary repair was associated with improved outcome compared with a two-stage approach [20]. However, the impact of this more aggressive surgical approach on late survival or recurrent right ventricular outflow tract (RVOT) pathology is still unclear.

To clarify some of these issues, we report our surgical experience during three decades with TOF patients, with emphasis on the evolution to earlier repair and the influence this has had on early outcome and recurrent right heart obstruction.

Material and Methods

Patients

The medical records of all patients who underwent operation for TOF at the Oklahoma University Health Sciences Center between 1971 and October 1997 were reviewed. These included patients with classic TOF with pulmonary stenosis (TOF-PS) and those with associated pulmonary atresia (TOF-PA), but excludes other complex associated pathologic processes such as absent pulmonary valve syndrome or common atrioventricular canal.

Presented at the Forty-fourth Annual Meeting of the Southern Thoracic Surgical Association, Naples, FL, Nov 6–8, 1997.

Address reprint requests to Dr Knott-Craig, University of Oklahoma Health Sciences Center, PO Box 26901, Oklahoma City, OK 73190.

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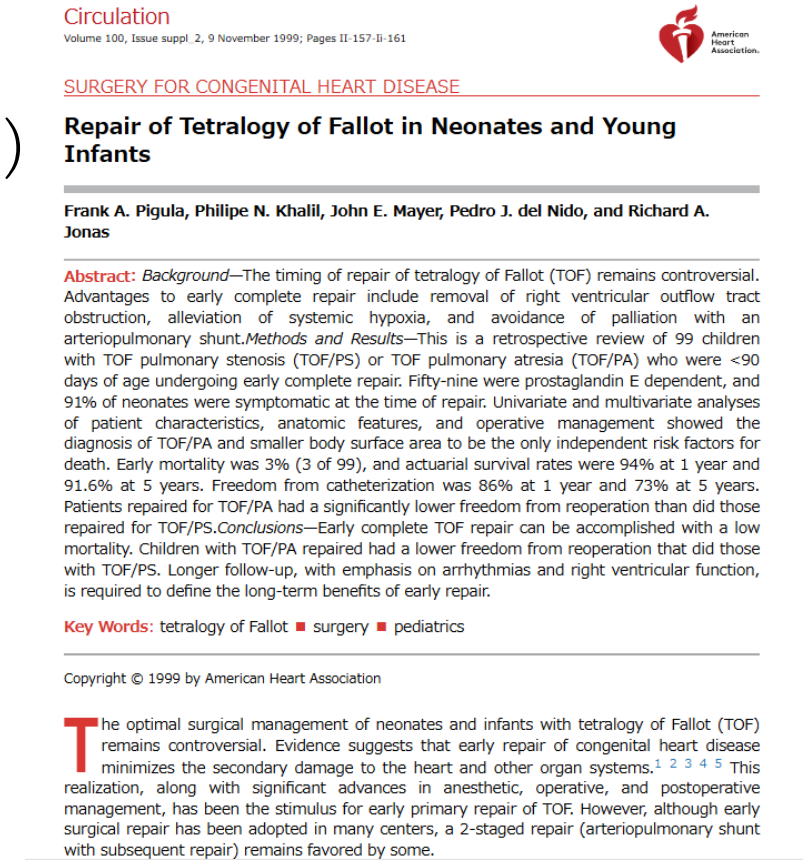
Ann Thorac Surg. 1998;66:506-11.

Early and late outcomes

- Pigula et al 1999
  - 99 children (< 90 days) had early repair (1988 -96)
  - 70/99 with transannular patch (TAP)
  - 96/99 with transventricular VSD repair

  - 3% early mortality
  - 91.6% survival at 5 years
  - 73% freedom from re-intervention at 5 years



Circulation
Volume 100, Issue suppl 2, 9 November 1999; Pages II-157-Ii-161

SURGERY FOR CONGENITAL HEART DISEASE

Repair of Tetralogy of Fallot in Neonates and Young Infants

Frank A. Pigula, Philipe N. Khalil, John E. Mayer, Pedro J. del Nido, and Richard A. Jonas

Abstract: *Background*—The timing of repair of tetralogy of Fallot (TOF) remains controversial. Advantages to early complete repair include removal of right ventricular outflow tract obstruction, alleviation of systemic hypoxia, and avoidance of palliation with an arteriopulmonary shunt.*Methods and Results*—This is a retrospective review of 99 children with TOF pulmonary stenosis (TOF/PS) or TOF pulmonary atresia (TOF/PA) who were <90 days of age undergoing early complete repair. Fifty-nine were prostaglandin E dependent, and 91% of neonates were symptomatic at the time of repair. Univariate and multivariate analyses of patient characteristics, anatomic features, and operative management showed the diagnosis of TOF/PA and smaller body surface area to be the only independent risk factors for death. Early mortality was 3% (3 of 99), and actuarial survival rates were 94% at 1 year and 91.6% at 5 years. Freedom from catheterization was 86% at 1 year and 73% at 5 years. Patients repaired for TOF/PA had a significantly lower freedom from reoperation than did those repaired for TOF/PS.*Conclusions*—Early complete TOF repair can be accomplished with a low mortality. Children with TOF/PA repaired had a lower freedom from reoperation that did those with TOF/PS. Longer follow-up, with emphasis on arrhythmias and right ventricular function, is required to define the long-term benefits of early repair.

Key Words: tetralogy of Fallot ■ surgery ■ pediatrics

The optimal surgical management of neonates and infants with tetralogy of Fallot (TOF) remains controversial. Evidence suggests that early repair of congenital heart disease minimizes the secondary damage to the heart and other organ systems.[1 2 3 4 5] This realization, along with significant advances in anesthetic, operative, and postoperative management, has been the stimulus for early primary repair of TOF. However, although early surgical repair has been adopted in many centers, a 2-staged repair (arteriopulmonary shunt with subsequent repair) remains favored by some.

Pigula F, et al. Circulation 1999;100: II-157-161.

Right ventriculotomy

- Belief that RVtomy made long-term RV dysfunction

- Shift the repair to trans-atrial and trans-pulmonary approach

J Thorac Cardiovasc Surg. 1992 Oct;104(4):917-23.

Evaluation of right ventricular function by regional wall motion analysis in patients after correction of tetralogy of Fallot. Comparison of transventricular and nontransventricular repairs.

Miura T[1], Nakano S, Shimazaki Y, Kobayashi J, Hirose H, Sano T, Matsuda H, Kawashima Y.

Author information

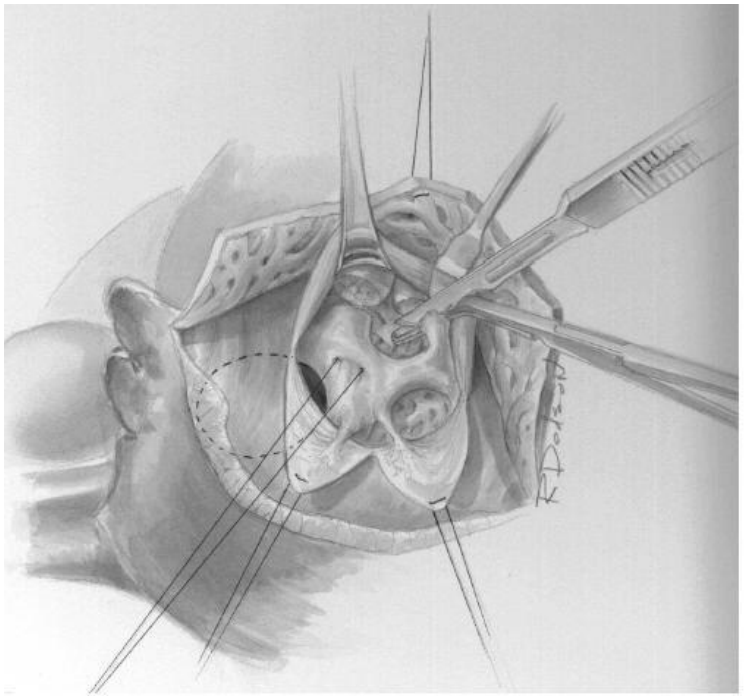
1 First Department of Surgery, Osaka University Medical School, Japan.

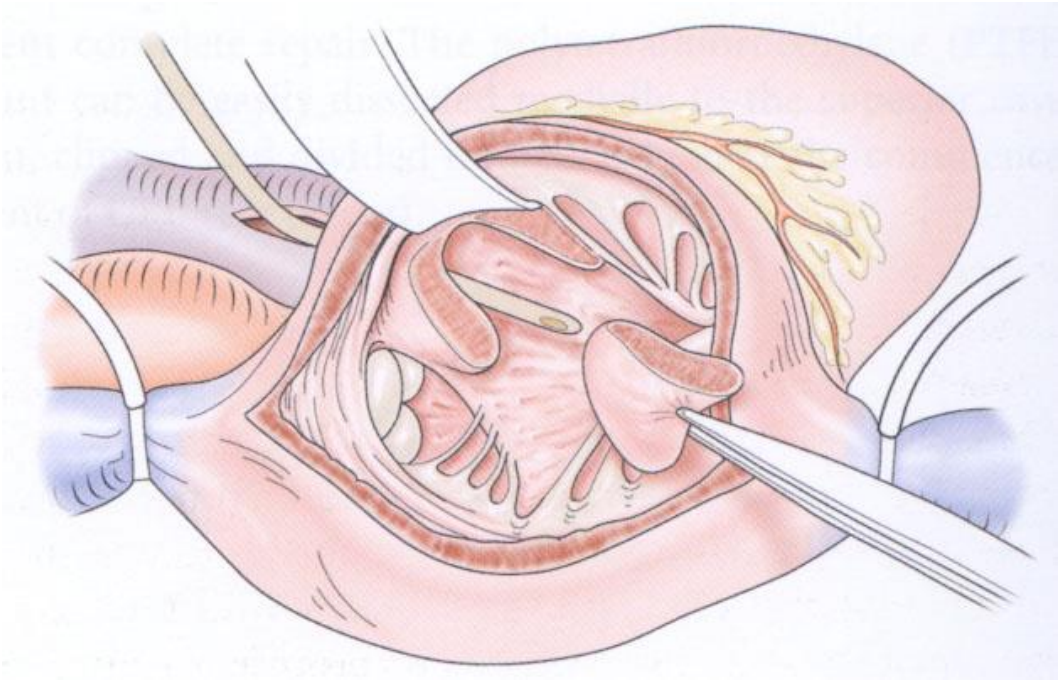
Abstract

Right ventricular function was assessed by regional wall motion analysis and by global function in 62 patients after repair for tetralogy of Fallot. Its relation to surgical procedures, with special attention to right ventriculotomy, was investigated. Patients were classified as follows: group Ia (n = 17), transpulmonary-transatrial repair without right ventriculotomy; group Ib (n = 22), transpulmonary-transatrial repair with

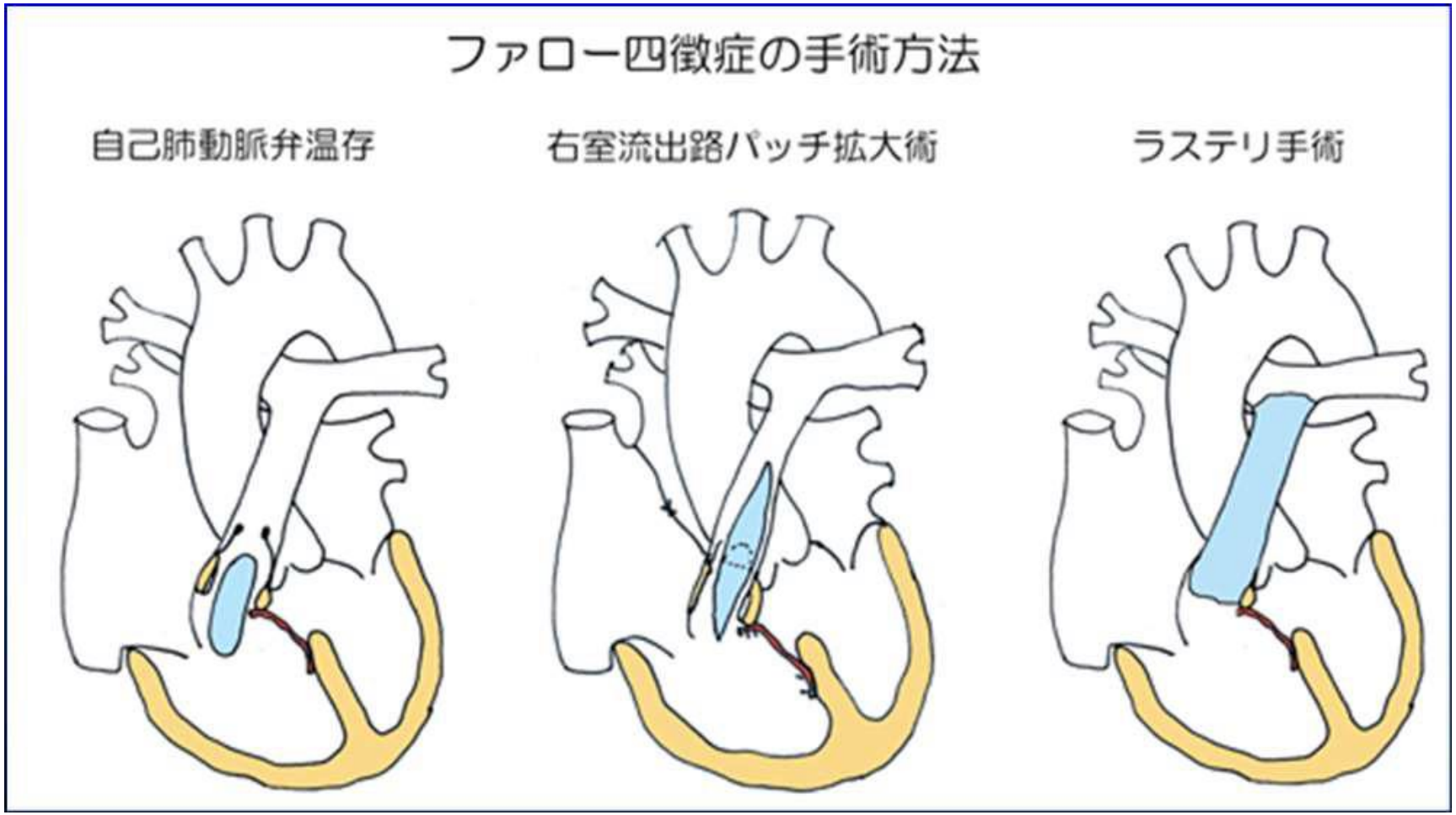
Pulmonary valve sparing TOF repair

- Trans-atrial and transpulmonary repair
  - Avoid or minimize ventriculotomy
    (limited to the outlet portion of RV or infundibulum)
  - Preserve pulmonary valve (z > -2)





Type of TOF repair



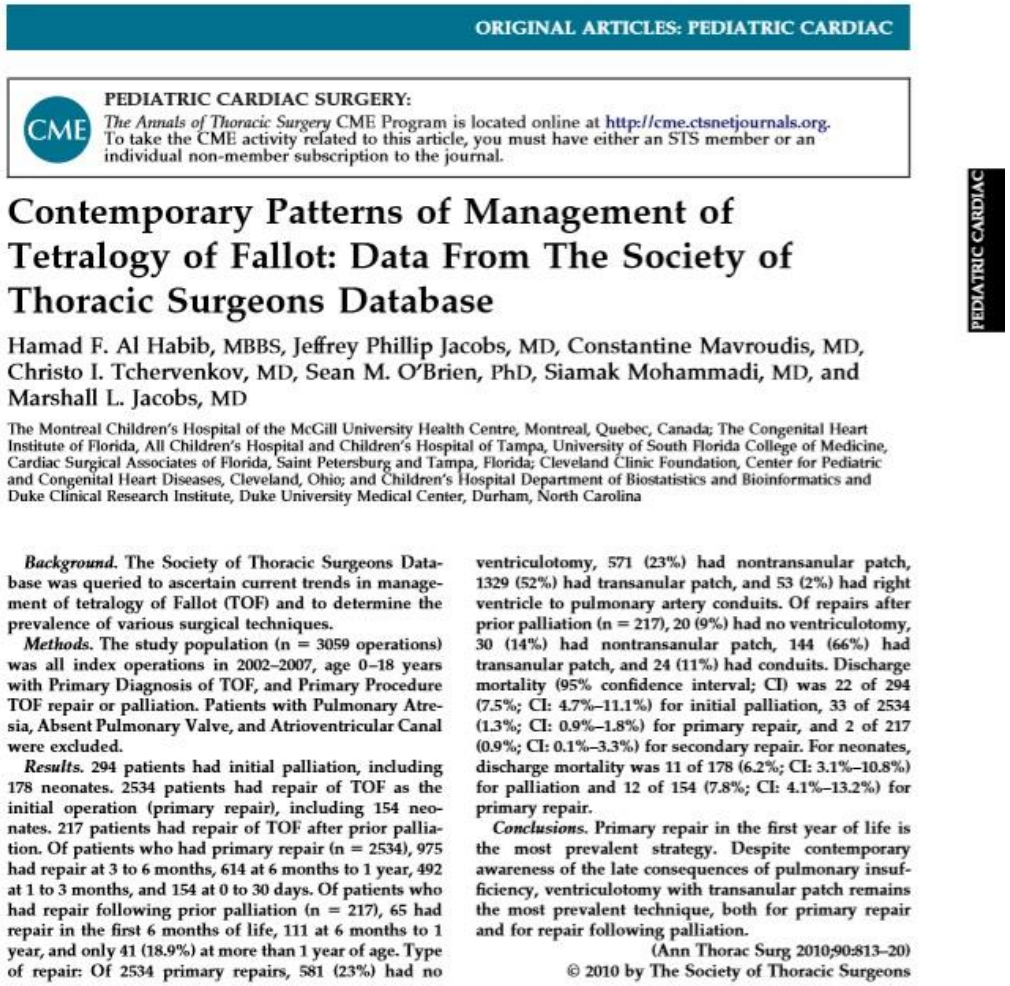
Pulmonary valve sparing
(RVMB resection +/- RVOT patch)

Transannular patch

RV-PA conduit

Current outcomes for TOF repair

- STS database study 1
  - 2002-2007
  - 3059 operations
  - < 18yo patients coded for TOF
    (exclude TOF/PA)
  - 294 had initial shunt
    (178 neonates)
  - 2534 primary complete repair
  - 217 repair after initial palliation



ORIGINAL ARTICLES: PEDIATRIC CARDIAC

Contemporary Patterns of Management of Tetralogy of Fallot: Data From The Society of Thoracic Surgeons Database

Hamad F. Al Habib, MBBS, Jeffrey Phillip Jacobs, MD, Constantine Mavroudis, MD, Christo I. Tchervenkov, MD, Sean M. O'Brien, PhD, Siamak Mohammadi, MD, and Marshall L. Jacobs, MD

The Montreal Children's Hospital of the McGill University Health Centre, Montreal, Quebec, Canada; The Congenital Heart Institute of Florida, All Children's Hospital and Children's Hospital of Tampa, University of South Florida College of Medicine, Cardiac Surgical Associates of Florida, Saint Petersburg and Tampa, Florida; Cleveland Clinic Foundation, Center for Pediatric and Congenital Heart Diseases, Cleveland, Ohio; and Children's Hospital Department of Biostatistics and Bioinformatics and Duke Clinical Research Institute, Duke University Medical Center, Durham, North Carolina

Background. The Society of Thoracic Surgeons Database was queried to ascertain current trends in management of tetralogy of Fallot (TOF) and to determine the prevalence of various surgical techniques.

Methods. The study population (n = 3059 operations) was all index operations in 2002–2007, age 0–18 years with Primary Diagnosis of TOF, and Primary Procedure TOF repair or palliation. Patients with Pulmonary Atresia, Absent Pulmonary Valve, and Atrioventricular Canal were excluded.

Results. 294 patients had initial palliation, including 178 neonates. 2534 patients had repair of TOF as the initial operation (primary repair), including 154 neonates. 217 patients had repair of TOF after prior palliation. Of patients who had primary repair (n = 2534), 975 had repair at 3 to 6 months, 614 at 6 months to 1 year, 492 at 1 to 3 months, and 154 at 0 to 30 days. Of patients who had repair following prior palliation (n = 217), 65 had repair in the first 6 months of life, 111 at 6 months to 1 year, and only 41 (18.9%) at more than 1 year of age. Type of repair: Of 2534 primary repairs, 581 (23%) had no ventriculotomy, 571 (23%) had nontransanular patch, 1329 (52%) had transanular patch, and 53 (2%) had right ventricle to pulmonary artery conduits. Of repairs after prior palliation (n = 217), 20 (9%) had no ventriculotomy, 30 (14%) had nontransanular patch, 144 (66%) had transanular patch, and 24 (11%) had conduits. Discharge mortality (95% confidence interval; CI) was 22 of 294 (7.5%; CI: 4.7%–11.1%) for initial palliation, 33 of 2534 (1.3%; CI: 0.9%–1.8%) for primary repair, and 2 of 217 (0.9%; CI: 0.1%–3.3%) for secondary repair. For neonates, discharge mortality was 11 of 178 (6.2%; CI: 3.1%–10.8%) for palliation and 12 of 154 (7.8%; CI: 4.1%–13.2%) for primary repair.

Conclusions. Primary repair in the first year of life is the most prevalent strategy. Despite contemporary awareness of the late consequences of pulmonary insufficiency, ventriculotomy with transanular patch remains the most prevalent technique, both for primary repair and for repair following palliation.

(Ann Thorac Surg 2010;90:813–20)

Al Habib HF, et al. Ann Thorac Surg. 2010;90:813-9.

Current outcomes for TOF repair

- STS database study 1
  - 2534 primary complete repair
    - 581 (23%) had no ventriculotomy
    - 571 (23%) had non-transanular patch
    - 1329 (52%) had transanular patch
    - 53 (2%) had RV-PA conduits.
  - 217 complete repairs with prior palliation
    - 20 (9%) had no ventriculotomy
    - 30 (14%) had non-transanular patch
    - 144 (66%) had transanular patch
    - 24 (11%) had RV-PA conduits.

Table 6. Tetralogy of Fallot Repair Without Previous Operation: Type of Repair

Type of TOF Repair	0–30 Days No.	>30 Days–3 Months No.	>3–6 Months No.	>6 Months–1 Year No.	>1–2 Years No.	>2–18 Years No.	All Ages No.
No ventriculotomy	9	91	237	172	44	28	581
Ventriculotomy, Nontransanular patch	13	82	222	169	37	48	571
Ventriculotomy, Transanular patch	126	312	507	266	58	60	1329
RV-PA conduit	6	7	9	7	6	18	53
All operations	154	492	975	614	145	154	2534

RV-PA = right ventricle-to-pulmonary artery; TOF = tetralogy of Fallot.

Al Habib HF, et al. Ann Thorac Surg. 2010;90:813-9.

Current outcomes for TOF repair

- STS database study 1

Table 8. Discharge Mortality Stratified by Type of Procedure and Age at Operation

TOF Procedure	0–30 Days No. (%)	>30 Days–3 Months No. (%)	>3–6 Months No. (%)	>6 Months–1 Year No. (%)	>1–2 Years No. (%)	>2–18 Years No. (%)	All Ages No. (%)
TOF palliation							
Without previous cardiac operation	11/178 (6.2)	7/89 (7.9)	2/16 (12.5)	2/6 (33)	0/1 (0)	0/4 (0)	22/294 (7.5)
After previous cardiac operation	0/4 (0)	0/6 (0)	0/1 (0)	0/2 (0)	0/1 (0)	0/0 (0)	0/14 (0)
TOF repair							
Without previous cardiac operation	12/154 (7.8)	8/492 (1.6)	6/975 (0.6)	4/614 (0.6)	1/145 (0.7)	2/154 (1.3)	33/2534 (1.3)
After previous cardiac operation	1/8 (12.5)	0/10 (0)	0/47 (0)	1/111 (0.9)	0/37 (0)	0/4 (0)	2/217 (0.9)
All operations	24/344 (7.0)	15/597 (2.5)	8/1039 (0.8)	7/733 (1.0)	1/184 (0.5)	2/162 (1.2)	57/3059 (1.9)

TOF = tetralogy of Fallot.

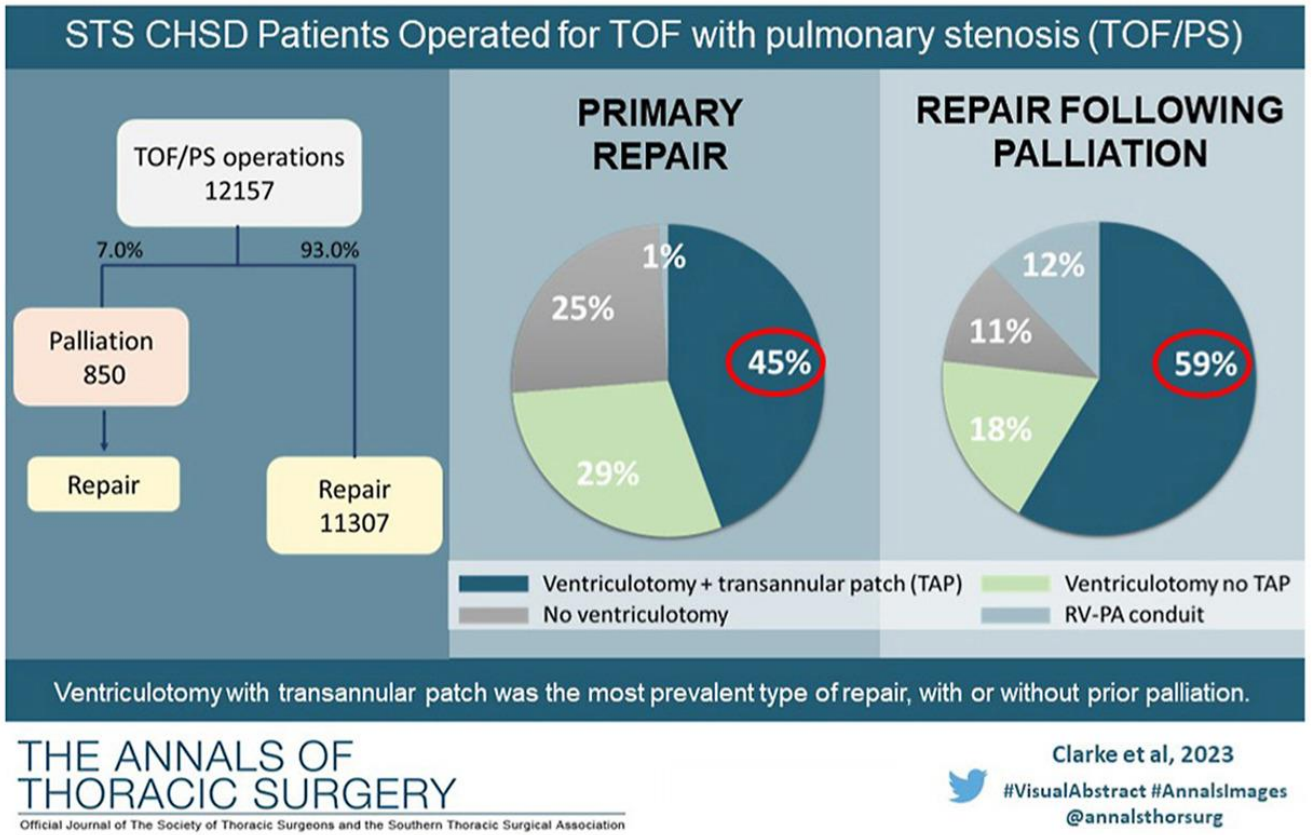
Al Habib HF, et al. Ann Thorac Surg. 2010;90:813-9.

Current outcomes for TOF repair

- STS database 2
  - 2010-2020
  - < 18yo patients coded for TOF (exclude TOF/PA)
  - 12157 index operation for TOF

  - Palliation: 7% in total cohort
    45% in neonates
    associated with higher preOp risk factor (50.1% vs 24.3%)
    higher mortality and morbidity (21.2% vs 7.46%)

Contemporary Patterns of Care in tetralogy of Fallot (TOF), 2010-2020



Clarke NS et al. Ann Thorac Surg. 2023;116:768-775.

Current outcomes for TOF repair in Japan

- National Clinical Database
  - Early mortality of TOF repair
    1.0% (2017-18)
    2.0% (2015-16)
  - Complications
    - Unplanned Reoperation 4.0%
    - Pacemaker implantation 0.7%
    - Wound infection 0.6%
    - Neurological deficit 0.4%

Table 1 Mortality and major complications of twenty major procedures of congenital heart surgery in Japan (2017 and 2018)

2017–2018	Total	Death (90 days or in-hospital)		(2015–2016)	Unplanned cardiac reoperation exclusive of reoperation for bleeding		Arrhythmia necessitating permanent pacemaker		Chylothorax		Wound infection (Mediastinitis/ deep wound infection)		Phrenic nerve injury		Neurological deficit persisting at discharge	
VSD repair	2,980	7	0.2%	0.3%	26	0.9%	8	0.3%	40	1.3%	14	0.5%	6	0.2%	3	0.1%
ASD repair	1,386	0	0.0%	0.1%	9	0.6%	6	0.4%	9	0.6%	2	0.1%	0	0.0%	3	0.2%
PDA closure	1,110	49	4.4%	3.9%	12	1.1%	1	0.1%	24	2.2%	1	0.1%	6	0.5%	2	0.2%
Systemic to pulmonary shunt	1,092	54	4.9%	4.7%	105	9.6%	9	0.8%	40	3.7%	9	0.8%	17	1.6%	4	0.4%
Pulmonary artery banding	1,023	47	4.6%	5.7%	51	5.0%	2	0.2%	27	2.6%	12	1.2%	6	0.6%	2	0.2%
TCPC (Fontan)	818	15	1.8%	2.6%	58	7.1%	16	2.0%	57	7.0%	21	2.6%	18	2.2%	6	0.7%
TOF repair	680	7	1.0%	2.0%	27	4.0%	5	0.7%	19	2.8%	4	0.6%	1	0.1%	3	0.4%
Bidirectional Glenn	647	17	2.6%	2.5%	45	7.0%	6	0.9%	52	8.0%	6	0.9%	19	2.9%	7	1.1%
CoA repair	440	13	3.0%	3.0%	23	5.2%	1	0.2%	31	7.0%	5	1.1%	11	2.5%	2	0.5%
CoA complex repair	223	5	2.2%	2.4%	12	5.4%	2	0.9%	15	6.7%	2	0.9%	11	4.9%	1	0.4%
Valve replacement (PVR)	387	2	0.5%	1.2%	6	1.6%	6	1.6%	1	0.3%	3	0.8%	4	1.0%	4	1.0%
TAPVC repair	359	40	11.1%	13.8%	41	11.4%	1	0.3%	39	10.9%	3	0.8%	11	3.1%	2	0.6%
Complete AVSD repair	349	7	2.0%	2.7%	24	6.9%	5	1.4%	41	11.7%	8	2.3%	1	0.3%	4	1.1%
Rastelli operation	268	6	2.2%	2.1%	14	5.2%	2	0.7%	16	6.0%	2	0.7%	6	2.2%	2	0.7%
Mitral valvuloplasty	246	13	5.3%	8.4%	13	5.3%	4	1.6%	6	2.4%	3	1.2%	1	0.4%	3	1.2%
PA plasty	242	6	2.5%	3.2%	15	6.2%	1	0.4%	12	5.0%	1	0.4%	7	2.9%	1	0.4%
Norwood procedure	229	36	15.7%	16.0%	24	10.5%	3	1.3%	28	12.2%	8	3.5%	16	7.0%	4	1.7%
Arterial switch operation (ASO)	219	15	6.8%	6.4%	16	7.3%	1	0.5%	16	7.3%	0	0.0%	8	3.7%	0	0.0%
ASO with VSD repair	109	4	3.7%	9.8%	6	5.5%	0	0.0%	10	9.2%	0	0.0%	5	4.6%	0	0.0%
Pulmonary venous stenosis repair	126	12	9.5%	8.9%	14	11.1%	1	0.8%	11	8.7%	4	3.2%	7	5.6%	3	2.4%

VSD, ventricular septal defect ; ASD, atrial septal defect ; PDA, patent ductus arteriosus ; TCPC, total cavopulmonary connection ; TOF, tetralogy of Fallot ; CoA, coarctation of aorta ; TAPVC, total anomalous pulmonary venous connection ; AVSD, atrioventricular septal defect ; PA, pulmonary artery ; PAVSD, pulmonary atresia and ventricular septal defect.

Hirata. 日心外会誌 2020;49:151-15

When is the good time for TOF repair?

- How about neonatal TOF repair??

- Pros
  - Actuarial survival: 93% at 5 years
  - Free from re-Op: 89% at 1 year and 58% at 5 years
  - No neurological sequelae
  - 6.8 +/- 7 days mechanical ventilation

Hirsch JC et al. Ann Surg. 2000;232:508-14.

- Cons
  - Almost same mortality with shunt or repair
  - Individualized approach

Kanter KR et al. Ann Thorac Surg 2010;89:858-63.

When is the good time for TOF repair?

- PHIS database study
  - 2004-2010
  - 39 centers, 4968 operations
  - < 19yo patients

  - Discharge mortality of TOF
    → 1.3% (all cohort)
    6.4% (neonate)

※neonate group had higher morbidity

Timing of complete repair of non–ductal-dependent tetralogy of Fallot and short-term postoperative outcomes, a multicenter analysis

Matthew B. Steiner, MD,[a] Xinyu Tang, PhD,[b] Jeffrey M. Gossett, MS,[b] Sadia Malik, MD,[a] and Parthak Prodhan, MBBS[a,c]

Objective: There is cross-center variability with regard to timing repair of non–ductal-dependent tetralogy of Fallot (TOF). We hypothesized that earlier repair in the neonatal period is associated with increased mortality and morbidity.

Methods: This was a retrospective analysis of the Pediatric Health Information System of tetralogy of Fallot patients undergoing complete repair from 2004 through 2010 between the ages of 1 day to younger than 19 years. Patients with pulmonary valve atresia, those who received prostaglandin during hospital admission, and those who underwent prior shunt palliation were excluded.

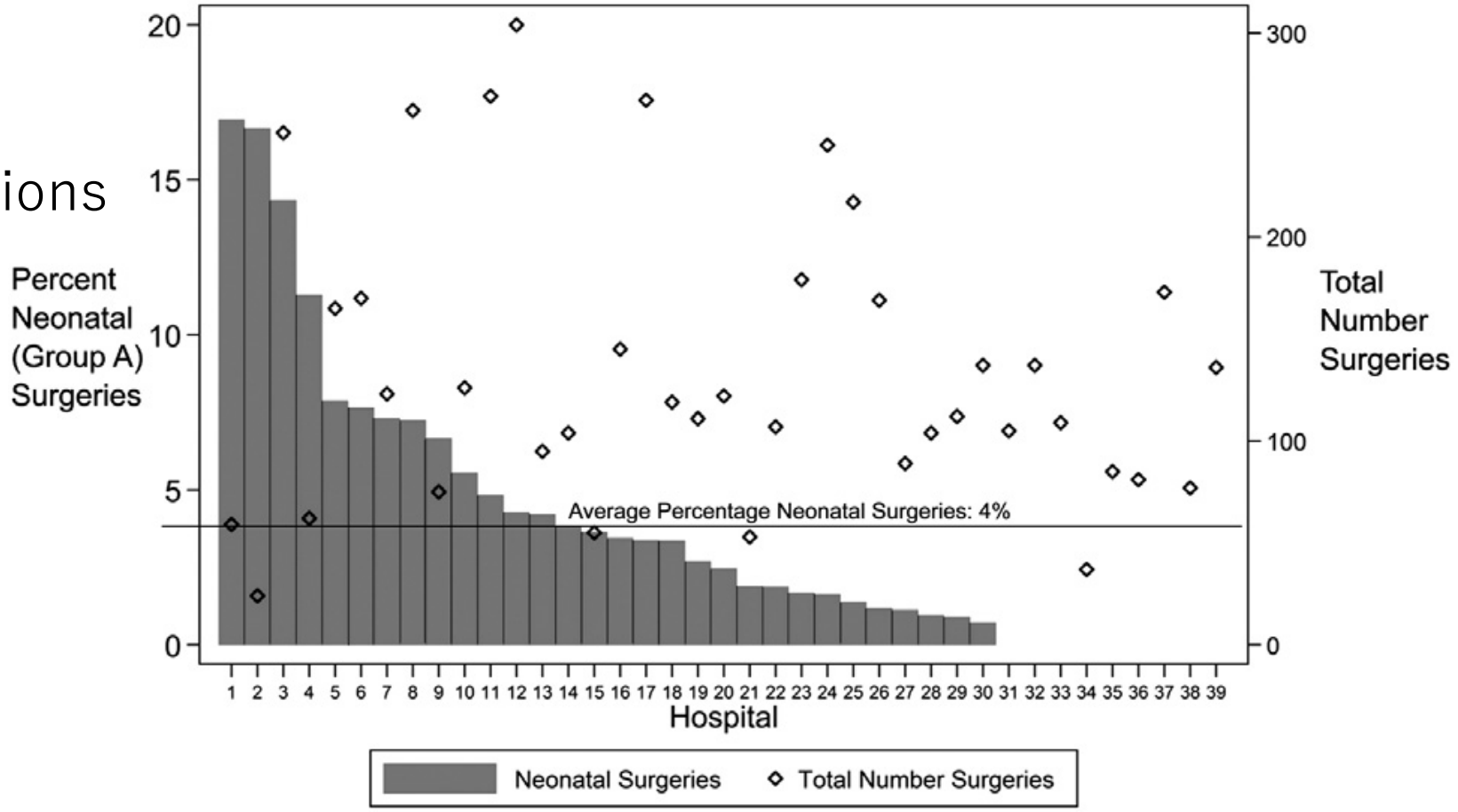
Results: A total of 4698 patients met our inclusion criteria, of whom 202 were younger than 30 days old (group A), 861 were 31 to 90 days old (group B), 1796 were 91 to 180 days old (group C), and 1839 were older than 180 days (group D). In-hospital mortality, intensive care unit length of stay, and total hospital length of stay were significantly higher in group A. Patients in group A had a significantly increased postoperative requirement for mechanical ventilation, intravenous blood pressure support, medical diuresis, extracorporeal membrane oxygenation, gastrostomy tube insertion, heart catheterization, and surgical revision. Significant institutional variability was noted for timing of TOF complete repair, with one third of the centers performing 75% of the repairs at younger than 30 days old. The institutional approach to timing TOF complete repair showed no relation to surgical volume.

Conclusions: Across all centers analyzed, primary neonatal elective TOF repair (<30 days of age) is associated with significantly higher postoperative in-hospital morbidity and mortality, although a few centers have shown an ability to use this strategy with excellent survivability. (J Thorac Cardiovasc Surg 2014;147:1299-305)

Steiner MB, et al. JTCVS 2014;147:1299-305.

When is the good time for TOF repair?

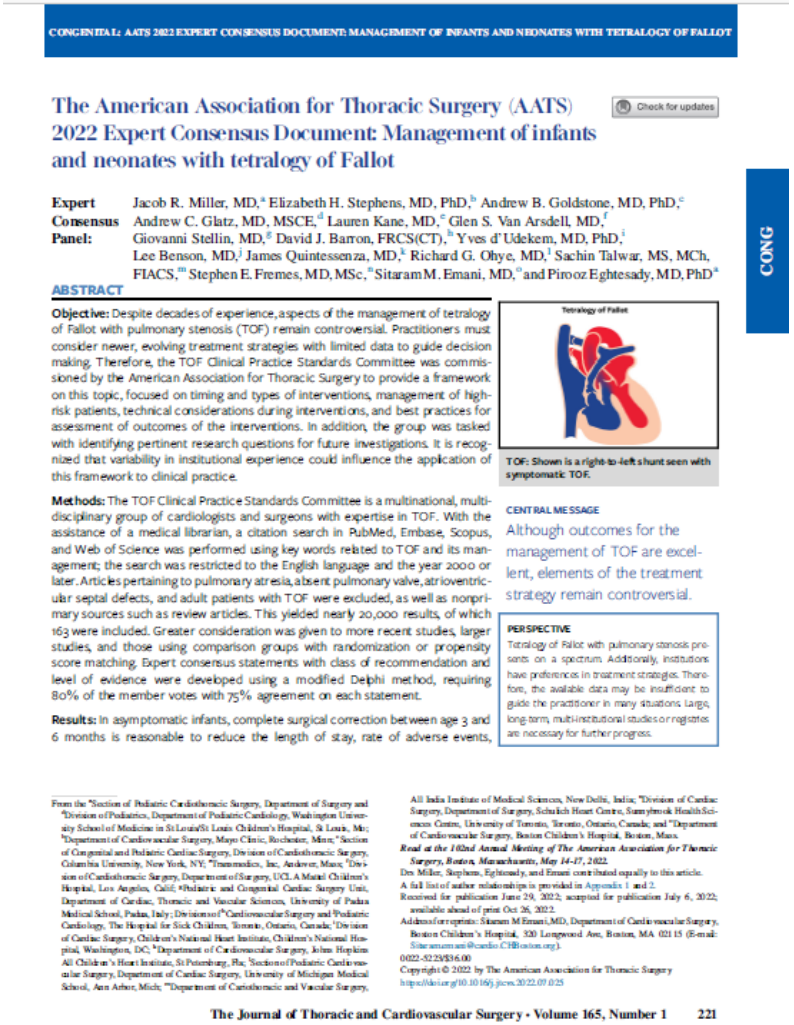
- PHIS database study
  - 2004-2010
  - 39 centers, 4968 operations
  - < 19yo patients
  - Significant variation in percentage of neonatal TOF repair



Steiner MB, et al. JTCVS 2014;147:1299-305.

When is the good time for TOF repair?

- North American consensus
  - Expert consensus panel
  - 163 papers reviewed
  - >75% expertise agreement on each statement



CONGENITAL: AATS 2022 EXPERT CONSENSUS DOCUMENT: MANAGEMENT OF INFANTS AND NEONATES WITH TETRALOGY OF FALLOT

The American Association for Thoracic Surgery (AATS) 2022 Expert Consensus Document: Management of infants and neonates with tetralogy of Fallot

Check for updates

Expert Consensus Panel: Jacob R. Miller, MD,[a] Elizabeth H. Stephens, MD, PhD,[b] Andrew B. Goldstone, MD, PhD,[c] Andrew C. Glatz, MD, MSCE,[d] Lauren Kane, MD,[e] Glen S. Van Arsdell, MD,[f] Giovanni Stellin, MD,[g] David J. Barron, FRCS(CT),[h] Yves d'Udekem, MD, PhD,[i] Lee Benson, MD,[j] James Quintessenza, MD,[k] Richard G. Ohye, MD,[l] Sachin Talwar, MS, MCh, FIACS,[m] Stephen E. Fremes, MD, MSc,[n] Sitaram M. Emani, MD,[o] and Pirooz Eghtesady, MD, PhD[a]

CONG

ABSTRACT

Objective: Despite decades of experience, aspects of the management of tetralogy of Fallot with pulmonary stenosis (TOF) remain controversial. Practitioners must consider newer, evolving treatment strategies with limited data to guide decision making. Therefore, the TOF Clinical Practice Standards Committee was commissioned by the American Association for Thoracic Surgery to provide a framework on this topic, focused on timing and types of interventions, management of high-risk patients, technical considerations during interventions, and best practices for assessment of outcomes of the interventions. In addition, the group was tasked with identifying pertinent research questions for future investigations. It is recognized that variability in institutional experience could influence the application of this framework to clinical practice.

Methods: The TOF Clinical Practice Standards Committee is a multinational, multidisciplinary group of cardiologists and surgeons with expertise in TOF. With the assistance of a medical librarian, a citation search in PubMed, Embase, Scopus, and Web of Science was performed using key words related to TOF and its management; the search was restricted to the English language and the year 2000 or later. Articles pertaining to pulmonary atresia, absent pulmonary valve, atrioventricular septal defects, and adult patients with TOF were excluded, as well as nonprimary sources such as review articles. This yielded nearly 20,000 results, of which 163 were included. Greater consideration was given to more recent studies, larger studies, and those using comparison groups with randomization or propensity score matching. Expert consensus statements with class of recommendation and level of evidence were developed using a modified Delphi method, requiring 80% of the member votes with 75% agreement on each statement.

Results: In asymptomatic infants, complete surgical correction between age 3 and 6 months is reasonable to reduce the length of stay, rate of adverse events,

TOF: Shown is a right-to-left shunt seen with symptomatic TOF.

CENTRAL MESSAGE

Although outcomes for the management of TOF are excellent, elements of the treatment strategy remain controversial.

PERSPECTIVE

Tetralogy of Fallot with pulmonary stenosis presents on a spectrum. Additionally, institutions have preferences in treatment strategies. Therefore, the available data may be insufficient to guide the practitioner in many situations. Large, long-term, multi-institutional studies or registries are necessary for further progress.

From the [a]Section of Pediatric Cardiothoracic Surgery, Department of Surgery and [b]Division of Pediatrics, Department of Pediatric Cardiology, Washington University School of Medicine in St Louis/St Louis Children's Hospital, St Louis, Mo; [c]Department of Cardiovascular Surgery, Mayo Clinic, Rochester, Minn; [d]Section of Congenital and Pediatric Cardiac Surgery, Division of Cardiothoracic Surgery, Columbia University, New York, NY; [e]Tenmedics, Inc, Andover, Mass; [f]Division of Cardiothoracic Surgery, Department of Surgery, UCL A Mattel Children's Hospital, Los Angeles, Calif; [g]Pediatric and Congenital Cardiac Surgery Unit, Department of Cardiac, Thoracic and Vascular Sciences, University of Padua Medical School, Padua, Italy; Division of [h]Cardiovascular Surgery and [i]Pediatric Cardiology, The Hospital for Sick Children, Toronto, Ontario, Canada; [j]Division of Cardiac Surgery, Children's National Heart Institute, Children's National Hospital, Washington, DC; [k]Department of Cardiovascular Surgery, Johns Hopkins All Children's Heart Institute, St Petersburg, Fla; [l]Section of Pediatric Cardiovascular Surgery, Department of Cardiac Surgery, University of Michigan Medical School, Ann Arbor, Mich; [m]Department of Cardiothoracic and Vascular Surgery, All India Institute of Medical Sciences, New Delhi, India; [n]Division of Cardiac Surgery, Department of Surgery, Schulich Heart Centre, Sunnybrook Health Sciences Centre, University of Toronto, Toronto, Ontario, Canada; and [o]Department of Cardiovascular Surgery, Boston Children's Hospital, Boston, Mass.

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Drs Miller, Stephens, Eghtesady, and Emani contributed equally to this article.

A full list of author relationships is provided in Appendix 1 and 2.

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Address for reprints: Sitaram M Emani, MD, Department of Cardiovascular Surgery, Boston Children's Hospital, 320 Longwood Ave, Boston, MA 02115 (E-mail: Sitaram.emani@cardio.CHBoston.org).

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J Thorac Cardiovasc Surg 2023;165:221-50.

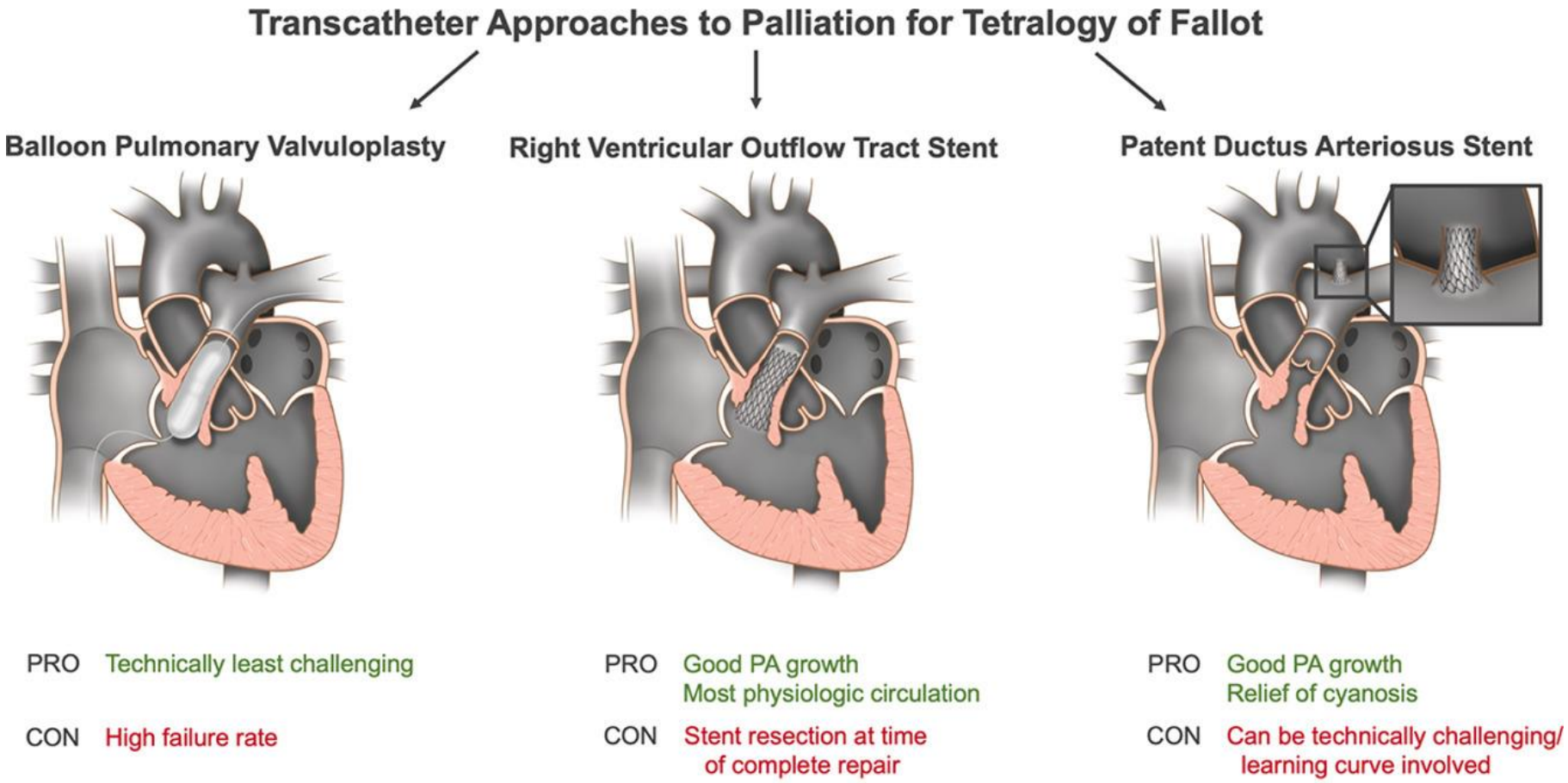
When is the good time for TOF repair?

Recommendation regarding elective complete surgical correction

Elective	COR	LOE
For patients with tetralogy of Fallot undergoing elective complete surgical correction, repair between ages 3 and 6 months is reasonable	IIa	B-NR
In standard-risk symptomatic neonates with TOF, both palliative procedures or complete surgical correction are reasonable at a center with demonstrated expertise.	IIa	B-NR

J Thorac Cardiovasc Surg 2023;165:221-50.

Catheter-based initial palliation?



When undergoing transcatheter palliation for Tetralogy of Fallot, r**ight ventricular outflow tract stenting and patent ductus arteriosus stenting** are preferred over balloon pulmonary valvuloplasty.

Qureshi AM, et al. Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu. 2022:25:48-57.

When to spare the pulmonary valve?

TABLE 1. Transannular patch (*TAP*) versus valve-sparing *z* scores

Reference	TAP *z* score	Valve-sparing *z* score
Lee and colleagues[86]	–2.7 ± 1.4	–0.8 ± 1.7
Mahajan and colleagues[49]	–2.4 ± 0.6	–1.9 ± 0.7
Bove and colleagues[87]	TAP: –3.26 ± 0.95 Infundibular sparing: –2.84 ± 0.75	–0.9 ± 1.15 (post-repair 0.34 ± 0.94)
Sen and colleagues[88]	–3.1 (range 0.8 to –9.0)	–1.4 (range 1.8 to –5.5)
Lozano-Balserio and colleagues[89]		Balloon dilation: –2.3 (range –1.3 to –4.5)
Robinson and colleagues[90]	–2.8 ± 1.0	No adjunct: –0.8 ± 1.2 Hegar dilation: –1.4 ± 1.1 Balloon dilation: –2.4 ± 1.0
Hofferberth and colleagues[91]		Balloon dilation: –2.2 (range –2.4 to –1.8)
Stephens and colleagues[92]	–3.1 (IQR –2.9 to –4.1)	–2.0 (IQR –2.4 to –1.8) "Ideal result" –1.2 ± 1.4
Hickey and colleagues[93]	–6.9	–4.5, post-repair successful –1.5 (range –2.6 to –1.0)
Vida and colleagues[94]	TAP with valve reconstruction: –3.35 (range –1.54 to –5.62)	Balloon dilation: –2.95 (range –0.95 to –4.06)
Choi and colleagues[95]	–2.5 ± 1.5	–0.9 ± 1.3
Zhao and colleagues[96]	–3.6 (range –2.6 to –5.3)	–1.5 (range –0.4 to –2.9)
Kasturi and colleagues[97]	–2.9 ± 1.4	–1.1 ± 1.6
Borodinova and colleagues[98]	–3.9 ± 2	–3.0 ± 1.9 (post-repair –1.0 ± 0.9)

IQR, Interquartile range.

For patients with TOF undergoing elective complete surgical correction with a preoperative pulmonary valve annular *z* score >–2.5, it is reasonable to consider a valve-sparing approach.	IIa	B-NR

J Thorac Cardiovasc Surg 2023;165:221-50.

Pulmonary valve creation

- Conotruncal repair
- Monocusp patch

Conotruncal Repair of Tetralogy of Fallot

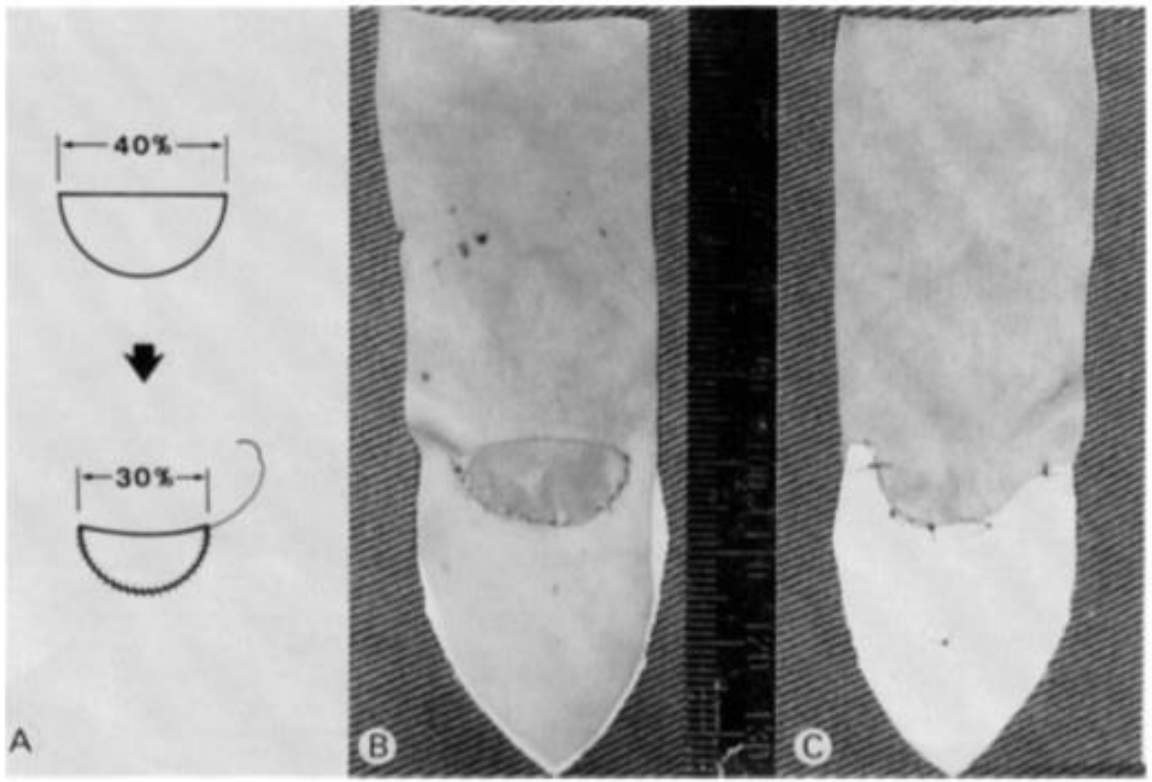
Hiromi Kurosawa, M.D., Yasuharu Imai, M.D., Makoto Nakazawa, M.D., Kazuo Momma, M.D., and Atsuyoshi Takao, M.D.

ABSTRACT **A repair within the con the right ventricle is introduced. It intracardiac repair of 30 consecutive pat of Fallot. The infundibular septum wa reduce the muscular outflow stenosis. dibuloplasty with a large monocusp stead of the tricuspid septal leaflet, th was employed as the suture line for pa lar septal defect (VSD) to avoid a cond residual VSD, and fixing of the tricuspi right ventricular (RV) to systemic art was 50.0 ± 14.6% (N = 26) and right atr ± 2.5 mm Hg (N = 26) one month a end-diastolic volume was 93.0 ± 30.5% before operation and 96.7 ± 29.0% of after operation in the same patients. that a conotruncal repair can maintain with low right atrial pressure and with volume.**

Approximately ten years ago, we dev ized patch infundibuloplasty proced Fallot in which a patch infundibulop the length of the right ventricle and dibulectomy are sufficient to relieve t (RV) outflow narrowing, while still p tion [1]. The ventricular septal defe guarded on its inferior margin by th trabecula septomarginalis, and has a that can be safely used for suture achieve goals of low right atrial p murmur, and no conduction disturb deavored to make the length of the plasty as short as possible and to use for patching the VSD. The repair is within the conotruncal portion of the 1). Here we discuss this new meth results.

Material and Methods

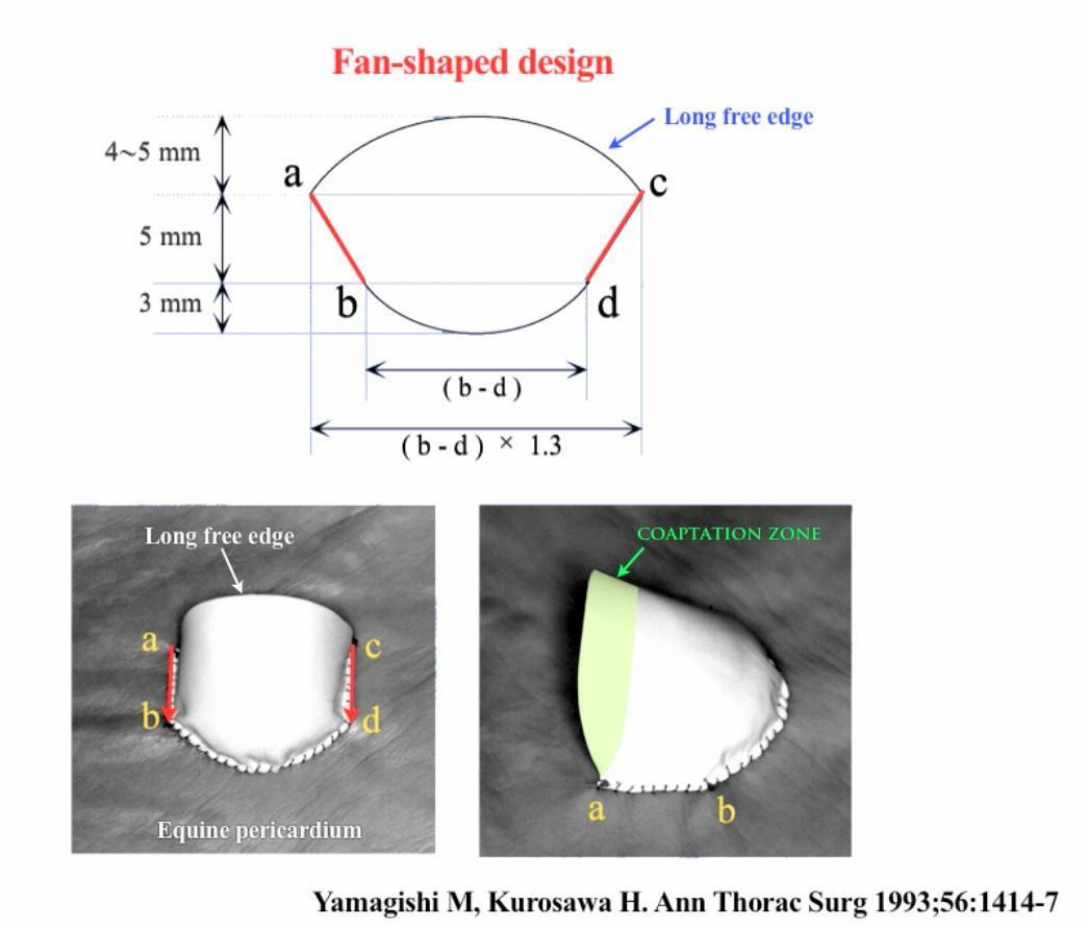
From 1985 to 1986, 30 consecutive pati



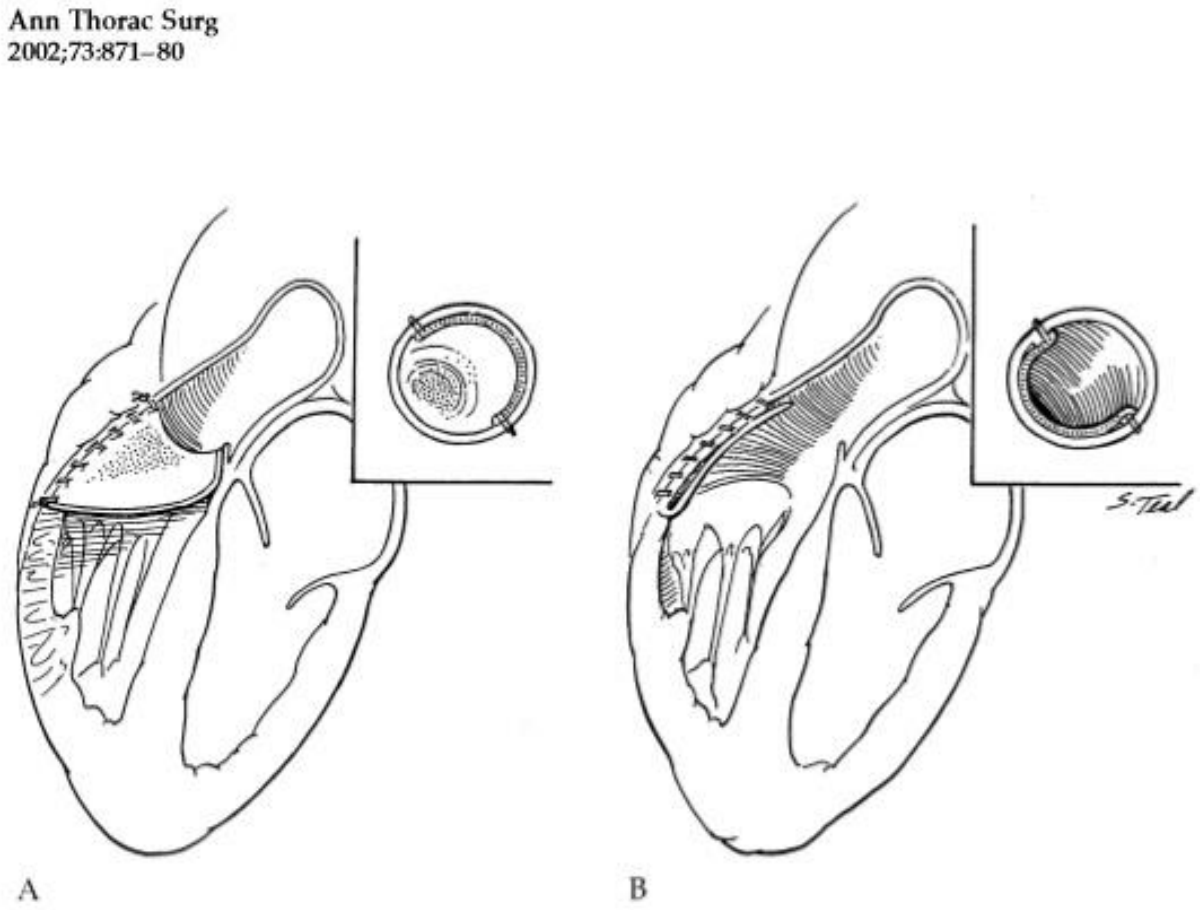
Ann Thorac Surg. 1988;45:661-6

Pulmonary valve creation

- ePTFE monocusp



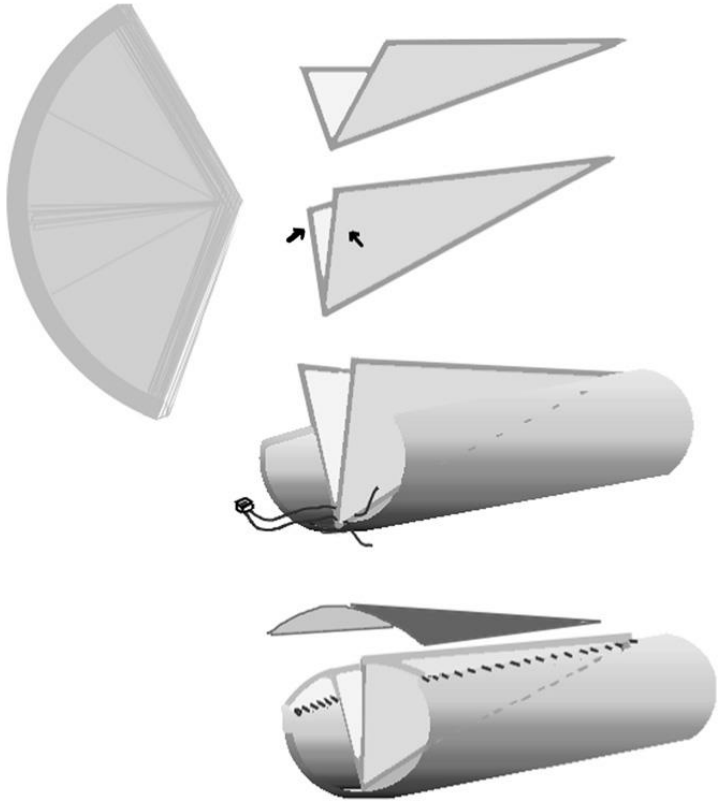
Yamagishi M, Kurosawa H. Ann Thorac Surg 1993;56:1414-7



Turrentin MW, et al. Ann Thorac Surg 2002;73:871–80.

Pulmonary valve creation

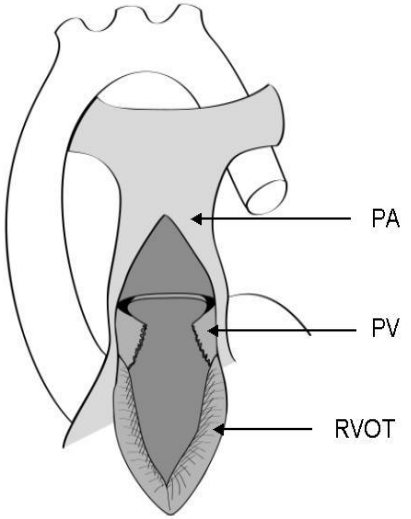
- ePTFE bi-leaflet valve (Nunn technique)

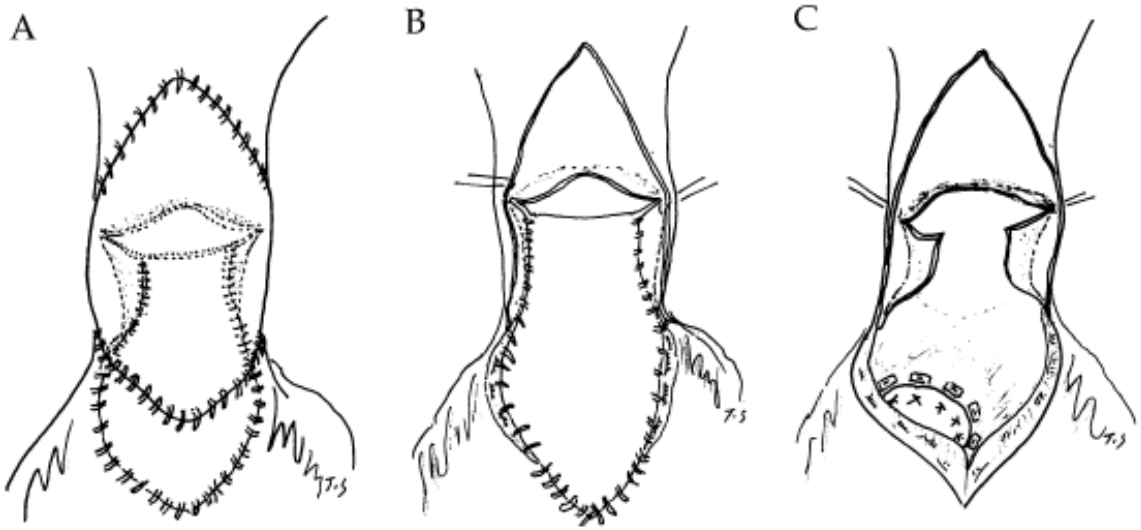


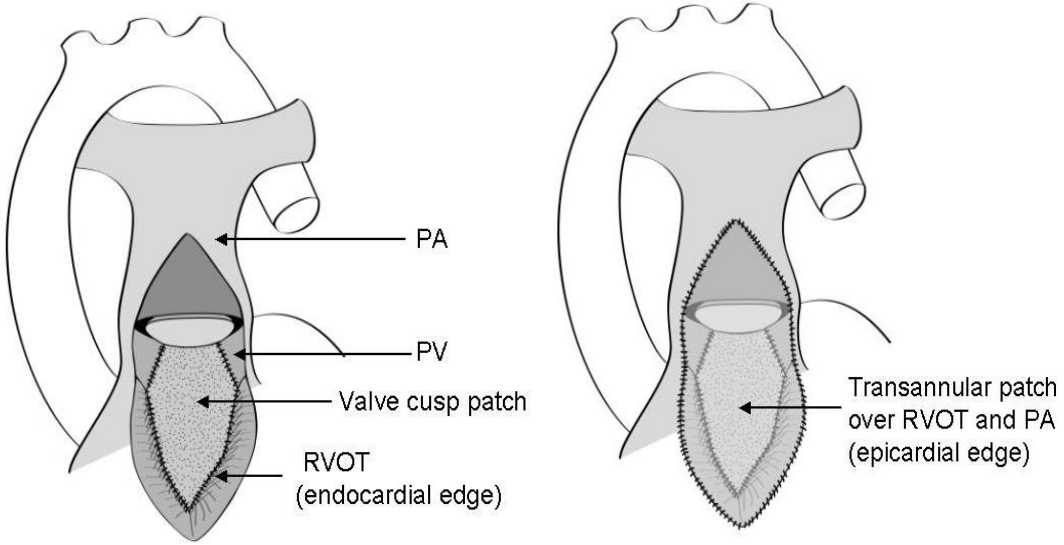
Nunn GR, et al. J Thorac Cardiovasc Surg 2008;136:290-7.

Pulmonary valve creation

- Pulmonary cusp augmentation
  - autologous pericardium





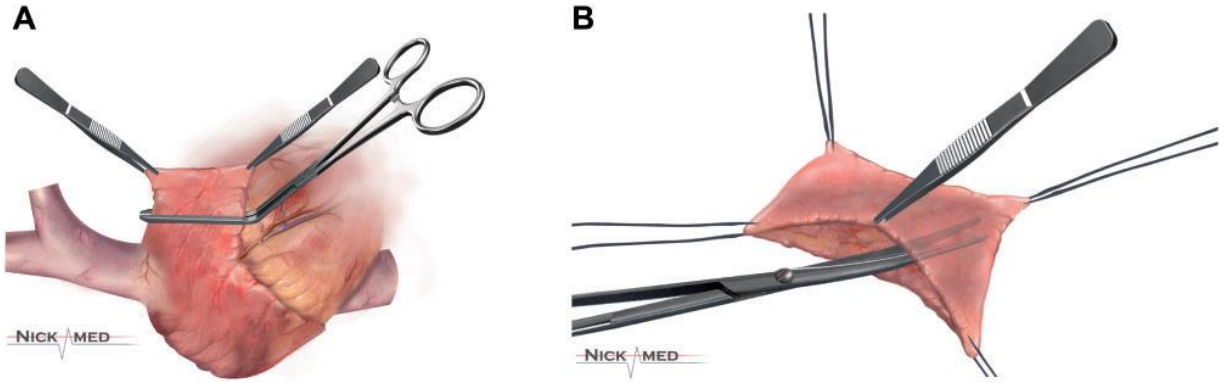


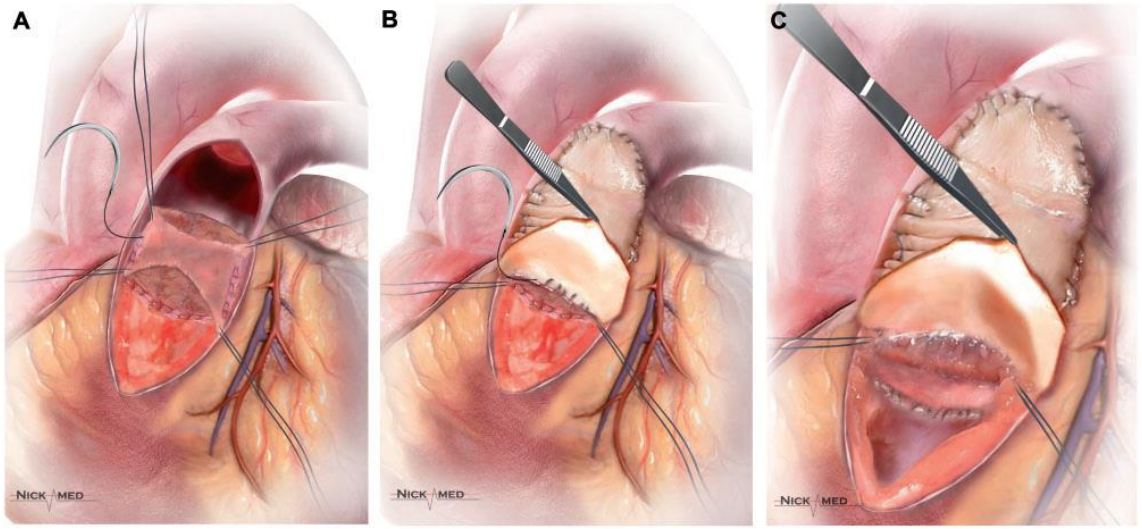
Sung SC. Ann Thorac Surg 2003;75:303–5.

Anagnostopoulos P et al. JTCVS. 2007;133:640-7.

Pulmonary valve creation

- Atrial appendage valve





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ORIGINAL ARTICLE

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Surgical repair of tetralogy of Fallot using autologous right atrial appendages: short- to mid-term results

Ahmadali Amirghofran[a], Fatemeh Edraki[b], Mohammadreza Edraki [c,*], Gholamhossein Ajami[c], Hamid Amoozgar[c], Hamid Mohammadi[c], Abbas Emaminia[d], Bahram Ghasemzadeh[a], Mohammad Borzuee[c], Farah Peiravian[e], Zahra Kheirandish[e], Nima Mehdizadegan [c], Mohammadreza Sabri[f], Sirus Cheriki[b] and Hamid Arabi[b]

[a] Cardiac Surgery Department, Shiraz University of Medical Sciences, Shiraz, Iran
[b] Shiraz University of Medical Sciences, Shiraz, Iran
[c] Cardiovascular and Neonatology Research Center, Shiraz University of Medical Sciences, Shiraz, Iran
[d] Inova Heart and Vascular Institute, Fall Church, VA, USA
[e] Pediatric Department, Faculty of Medicine, Islamic Azad University, Kazerun Branch, Kazerun, Iran
[f] Isfahan University of Medical Sciences, Isfahan, Iran

* Corresponding author. Pediatric Cardiology Ward, Namazi Hospital, Shiraz, Iran. Tel: +98-71-36125660; e-mail: edrakidr@yahoo.com (M. Edraki).

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Key question

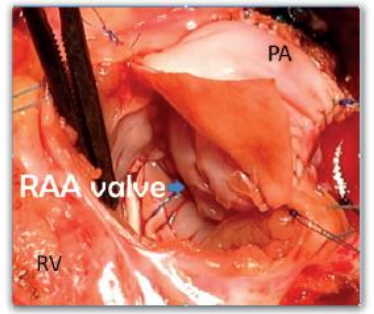
Can the right atrial appendage (RAA) be used to construct a bicuspid pulmonary valve in tetralogy of Fallot?

Key finding(s)

The RAA valve showed acceptable function with no/trivial pulmonary insufficiency in 81% of the cases in a 1-year follow-up.

Take-home message

The novel technique of RAA valve creation is safe, practical and reproducible with promising short- to mid-term outcomes.



Abstract

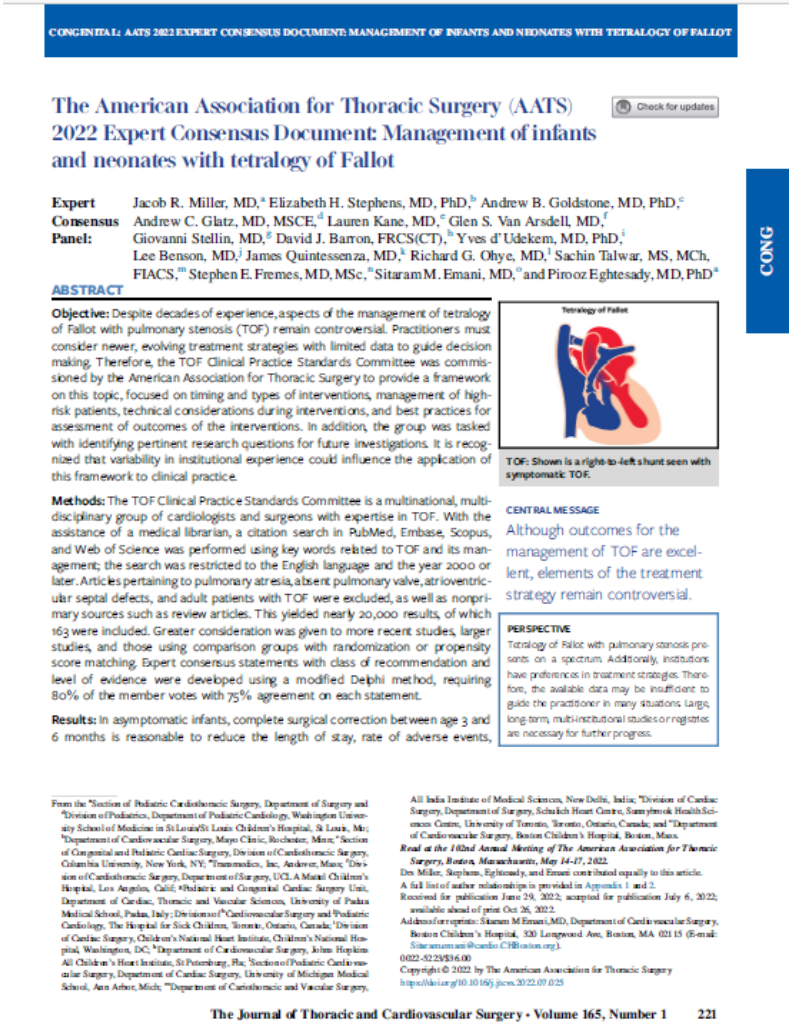
OBJECTIVES: The prevention of pulmonary insufficiency (PI) is a crucial part of the tetralogy of Fallot repair. Many techniques have been introduced to construct valves from different materials for the right ventricular outflow tract, including the most commonly constructed monocusp valves. We are introducing a new bicuspid valve made intraoperatively using the autologous right atrial appendage (RAA) to prevent PI in these patients.

Presented at the 33rd Annual Meeting of the European Association for Cardio-Thoracic Surgery, Lisbon, Portugal, 3–5 October 2019.

Eur J Cardiothorac Surg. 2021 Apr 13;59(3):697-704.

Which is the best technique for monocusp?

- North American consensus
  - From 163 papers
  - >75% expertise agreement on each statement



CONGENITAL: AATS 2022 EXPERT CONSENSUS DOCUMENT: MANAGEMENT OF INFANTS AND NEONATES WITH TETRALOGY OF FALLOT

The American Association for Thoracic Surgery (AATS) 2022 Expert Consensus Document: Management of infants and neonates with tetralogy of Fallot

Check for updates

Expert Consensus Panel: Jacob R. Miller, MD,[a] Elizabeth H. Stephens, MD, PhD,[b] Andrew B. Goldstone, MD, PhD,[c] Andrew C. Glatz, MD, MSCE,[d] Lauren Kane, MD,[e] Glen S. Van Arsdell, MD,[f] Giovanni Stellin, MD,[g] David J. Barron, FRCS(CT),[h] Yves d'Udekem, MD, PhD,[i] Lee Benson, MD,[j] James Quintessenza, MD,[k] Richard G. Ohye, MD,[l] Sachin Talwar, MS, MCh, FIACS,[m] Stephen E. Fremes, MD, MSc,[n] Sitaram M. Emani, MD,[o] and Pirooz Eghtesady, MD, PhD[a]

ABSTRACT

Objective: Despite decades of experience, aspects of the management of tetralogy of Fallot with pulmonary stenosis (TOF) remain controversial. Practitioners must consider newer, evolving treatment strategies with limited data to guide decision making. Therefore, the TOF Clinical Practice Standards Committee was commissioned by the American Association for Thoracic Surgery to provide a framework on this topic, focused on timing and types of interventions, management of high-risk patients, technical considerations during interventions, and best practices for assessment of outcomes of the interventions. In addition, the group was tasked with identifying pertinent research questions for future investigations. It is recognized that variability in institutional experience could influence the application of this framework to clinical practice.

Methods: The TOF Clinical Practice Standards Committee is a multinational, multidisciplinary group of cardiologists and surgeons with expertise in TOF. With the assistance of a medical librarian, a citation search in PubMed, Embase, Scopus, and Web of Science was performed using key words related to TOF and its management; the search was restricted to the English language and the year 2000 or later. Articles pertaining to pulmonary atresia, absent pulmonary valve, atrioventricular septal defects, and adult patients with TOF were excluded, as well as nonprimary sources such as review articles. This yielded nearly 20,000 results, of which 163 were included. Greater consideration was given to more recent studies, larger studies, and those using comparison groups with randomization or propensity score matching. Expert consensus statements with class of recommendation and level of evidence were developed using a modified Delphi method, requiring 80% of the member votes with 75% agreement on each statement.

Results: In asymptomatic infants, complete surgical correction between age 3 and 6 months is reasonable to reduce the length of stay, rate of adverse events,

Tetralogy of Fallot

TOF: Shown is a right-to-left shunt seen with symptomatic TOF.

CENTRAL MESSAGE

Although outcomes for the management of TOF are excellent, elements of the treatment strategy remain controversial.

PERSPECTIVE

Tetralogy of Fallot with pulmonary stenosis presents on a spectrum. Additionally, institutions have preferences in treatment strategies. Therefore, the available data may be insufficient to guide the practitioner in many situations. Large, long-term, multi-institutional studies or registries are necessary for further progress.

CONG

From the [a]Section of Pediatric Cardiothoracic Surgery, Department of Surgery and [b]Division of Pediatrics, Department of Pediatric Cardiology, Washington University School of Medicine in St Louis/St Louis Children's Hospital, St Louis, Mo; [c]Department of Cardiovascular Surgery, Mayo Clinic, Rochester, Minn; [d]Section of Congenital and Pediatric Cardiac Surgery, Division of Cardiothoracic Surgery, Columbia University, New York, NY; [e]Tenmedics, Inc, Andover, Mass; [f]Division of Cardiothoracic Surgery, Department of Surgery, UCL A Mattel Children's Hospital, Los Angeles, Calif; [g]Pediatric and Congenital Cardiac Surgery Unit, Department of Cardiac, Thoracic and Vascular Sciences, University of Padua Medical School, Padua, Italy; Division of [h]Cardiovascular Surgery and [i]Pediatric Cardiology, The Hospital for Sick Children, Toronto, Ontario, Canada; [j]Division of Cardiac Surgery, Children's National Heart Institute, Children's National Hospital, Washington, DC; [k]Department of Cardiovascular Surgery, Johns Hopkins All Children's Heart Institute, St Petersburg, Fla; [l]Section of Pediatric Cardiovascular Surgery, Department of Cardiac Surgery, University of Michigan Medical School, Ann Arbor, Mich; [m]Department of Cardiothoracic and Vascular Surgery, All India Institute of Medical Sciences, New Delhi, India; [n]Division of Cardiac Surgery, Department of Surgery, Schulich Heart Centre, Sunnybrook Health Sciences Centre, University of Toronto, Toronto, Ontario, Canada; and [o]Department of Cardiovascular Surgery, Boston Children's Hospital, Boston, Mass.

Read at the 102nd Annual Meeting of The American Association for Thoracic Surgery, Boston, Massachusetts, May 14-17, 2022.

Drs Miller, Stephens, Eghtesady, and Emani contributed equally to this article.

A full list of author relationships is provided in Appendix 1 and 2.

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Address for reprints: Sitaram M Emani, MD, Department of Cardiovascular Surgery, Boston Children's Hospital, 320 Longwood Ave, Boston, MA 02115 (E-mail: Sitaram.emani@cardio.CHBoston.org).

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J Thorac Cardiovasc Surg 2023;165:221-50.

Which is the best technique for monocusp?

- North American consensus
  - From 163 papers
  - >75% expertise agreement on each statement

TABLE 2. Valve reconstruction techniques

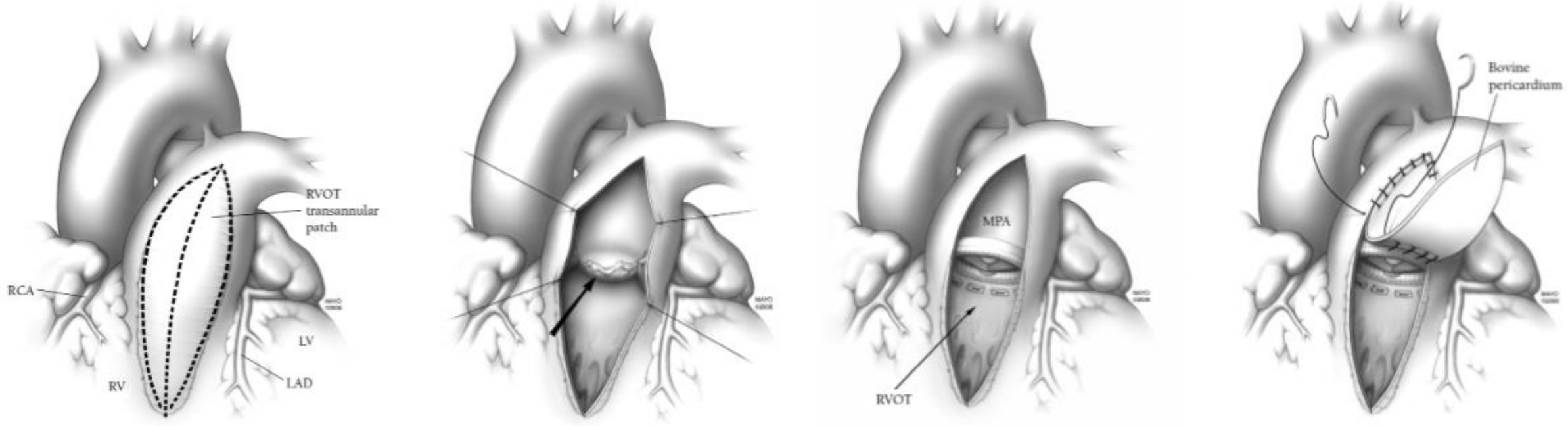
Reference	Year	n	Technique	Outcome
Patukale and colleagues[101]	2021	120	Leaflet augmentation: Various materials	Freedom from moderate or greater PI of 69% at 5 y and 30% at 10 y
Amirghofran and colleagues[102]	2021	21	RAA	RAA valve patients had shorter hospital LOS, similar PS, and less PI at 12 mo
Onan and colleagues[103]	2020	12	RAA	No PI in 11 and mild in 1 at 6 mo
Samadi and colleagues[104]*	2020	30	Monocusp: 0.1 mm PTFE	Compared to regular TAP, less rate of severe PI with monocusp
Vida and colleagues[105]	2019	12	Leaflet delamination and resuspension	None/mild PI in 10 and moderate in 2 at 2.8 y follow-up
Singh and colleagues[106]	2018	43	Monocusp: Various materials	Less PI but no improvement in clinical outcomes
Aydin and colleagues[107]	2018	15	Pericardial monocusp	Less PI and overall morbidity with monocusp than only TAP
Kumar and colleagues[108]	2016	171	Monocusp: 0.1 mm PTFE	At 10 y only 42 required monocusp; Severe PI in <25%; severe PS in <10%
Jang and colleagues[109]	2015	25	Monocusp: 0.1 mm PTFE	Prolonged the development of severe PI
Sasson and colleagues[110]	2012	74	Monocusp: 0.4 mm PTFE	Monocusp echo results and clinical outcomes (vent d, ICU LOS, chest tube drainage) were similar to those who underwent valve preservation; both better than with TAP alone
Pande and colleagues[111]	2010	16	Monocusp: Pericardium	Reduced PI at 1 y without difference in clinical outcomes
Gil-Jaurena and colleagues[112]	2010	21	Monocusp: 0.1 mm PTFE	Low rates of PI or PS on discharge echo
Park and colleagues[113]	2009	130	Monocusp: various materials	No influence on early mortality or risk of reoperation
Quintessenza and colleagues[114]	2009	126	Bicuspid: 0.6 then 0.1 mm PTFE	Only 6 of the valves failed, all 0.6 mm type failed due to calcified leaflets
Anagnostopoulos and colleagues[115]	2007	18	Cusp augmentation	Cusp augmentation group had shorter ventilation and inotrope dependence, shorter ICU LOS, and less PI at discharge
He and colleagues[116]	2006	9	Monocusp: Pericardium	Mild PI in 5 and minimal in 4 on discharge echo

n, Number of patients who underwent valve reconstructive technique; *PI*, pulmonary insufficiency; *RAA*, right atrial appendage; *LOS*, length of stay; *PS*, pulmonary stenosis; *PTFE*, polytetrafluoroethylene; *TAP*, transannular patch; *echo*, echocardiogram. *All studies are retrospective single institution studies with the exception of prospective randomized controlled trials.

J Thorac Cardiovasc Surg 2023;165:221-50.

Pulmonary valve replacement (PVR)

- (Redo) median sternotomy, regular cardiopulmonary bypass
- Under aortic cross-clamp (cardioplegic arrest) or Vf (electrical arrest)



Operative Techniques in Thoracic and Cardiovascular Surgery 2016

Long-term PVR - Indication?

- PVR to control PI
- RV muscle resection to relieve the obstruction

- Operative indications
  - To improve symptoms and avoid sudden death
    - RV dilatation
    - RV dysfunction
    - TR
    - QRS elongation
    - Arrhythmia

Table. Proposed Indications for PVR in Patients With Repaired TOF or Similar Physiology With Moderate or Severe PR (Regurgitation Fraction ≥25%)

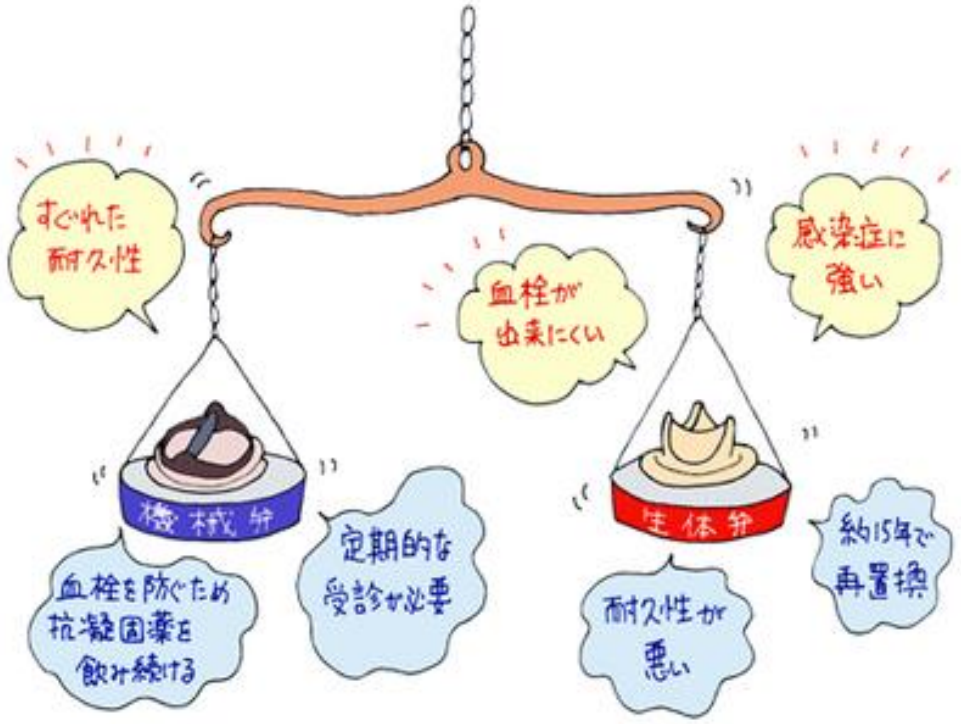
Indications	Supporting References
I. Asymptomatic patients with ≥2 of the following criteria:	
a. RV end-diastolic volume index >150 mL/m² or *z* score >4. In patients whose body surface area falls outside published normal data: RV/LV end-diastolic volume ratio >2	10, 12
b. RV end-systolic volume index >80 mL/m²	11, 13
c. RV ejection fraction <47%	11, 15, 16
d. LV ejection fraction <55%	11, 15, 16
e. Large RVOT aneurysm	17, 18
f. QRS duration >160 ms	11
g. Sustained tachyarrhythmia related to right-sided heart volume load	6
h. Other hemodynamically significant abnormalities:	
• RVOT obstruction with RV systolic pressure ≥0.7 systemic	19
• Severe branch pulmonary artery stenosis (<30% flow to affected lung) not amenable to transcatheter therapy	
• Greater than or equal to moderate tricuspid regurgitation	19
• Left-to-right shunt from residual atrial or ventricular septal defects with pulmonary-to-systemic flow ratio ≥1.5	19
• Severe aortic regurgitation	19
II. Symptomatic patients fulfilling ≥1 of the quantitative criteria detailed above. Examples of symptoms and signs include:	
a. Exercise intolerance not explained by extracardiac causes (eg, lung disease, musculoskeletal anomalies, genetic anomalies, obesity), with documentation by exercise testing with metabolic cart (≤70% predicted peak $\dot{V}o_2$ for age and sex not explained by chronotropic incompetence)	
b. Signs and symptoms of heart failure (eg, dyspnea with mild effort or at rest not explained by extracardiac causes, peripheral edema)	19
c. Syncope attributable to arrhythmia	
III. Special considerations:	
a. Because of higher risk of adverse clinical outcomes in patients who underwent TOF repair at ≥3 years of age, PVR may be considered if they fulfill ≥1 of the quantitative criteria in section I	16
b. Women with severe PR and RV dilatation or dysfunction may be at risk for pregnancy-related complications. Although no evidence is available to support benefit from prepregnancy PVR, the procedure may be considered if fulfilling ≥1 of the quantitative criteria in section I	20

LV indicates left ventricular; PR, pulmonary regurgitation; PVR, pulmonary valve replacement; RV, right ventricular; RVOT, right ventricular outflow tract; and TOF, tetralogy of Fallot.

Geva T. Circulation 2013;128:1855-7.

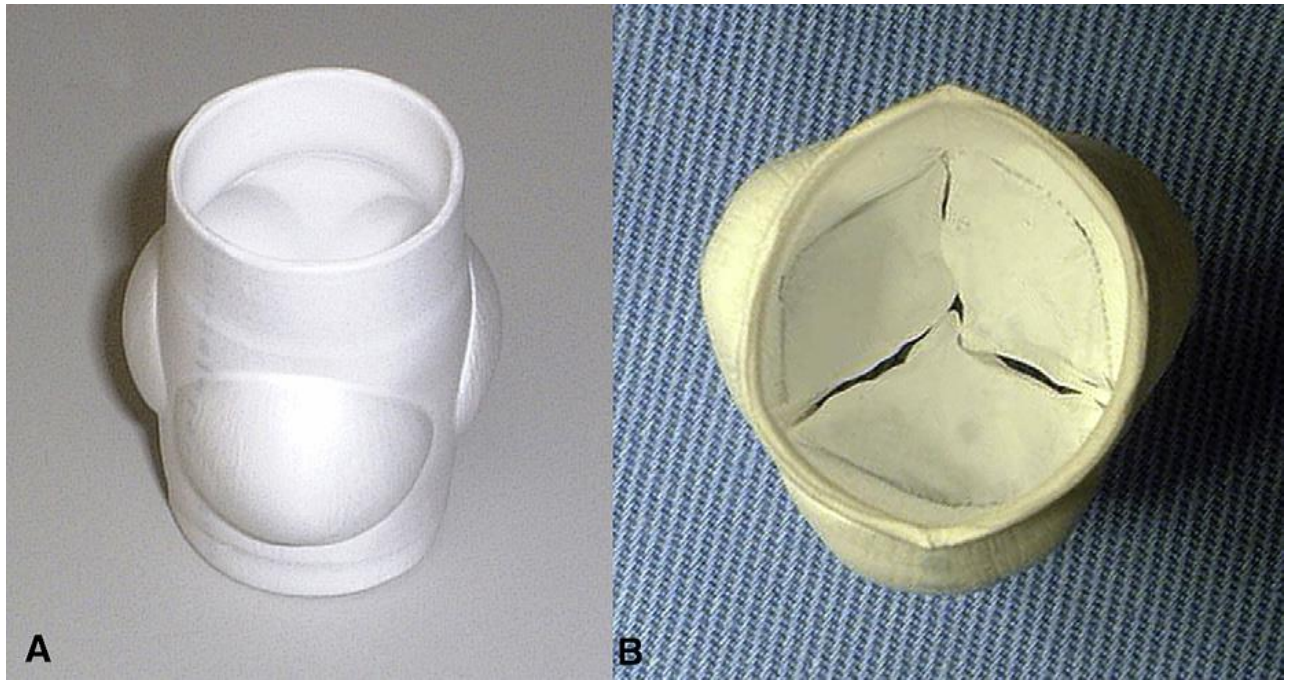
Pulmonary valve replacement (PVR)

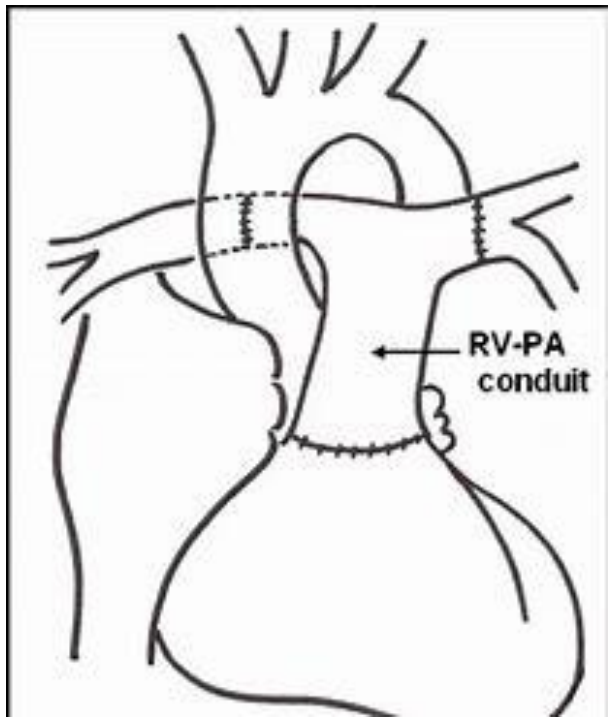
- Commonly PVR with bioprosthesis (rarely mechanical valve)



Pulmonary valve replacement (PVR)

- Only in some Japanese or Japanese-related overseas centers, PVR with valved ePTFE conduit (so-called “Yamagishi-valve”)

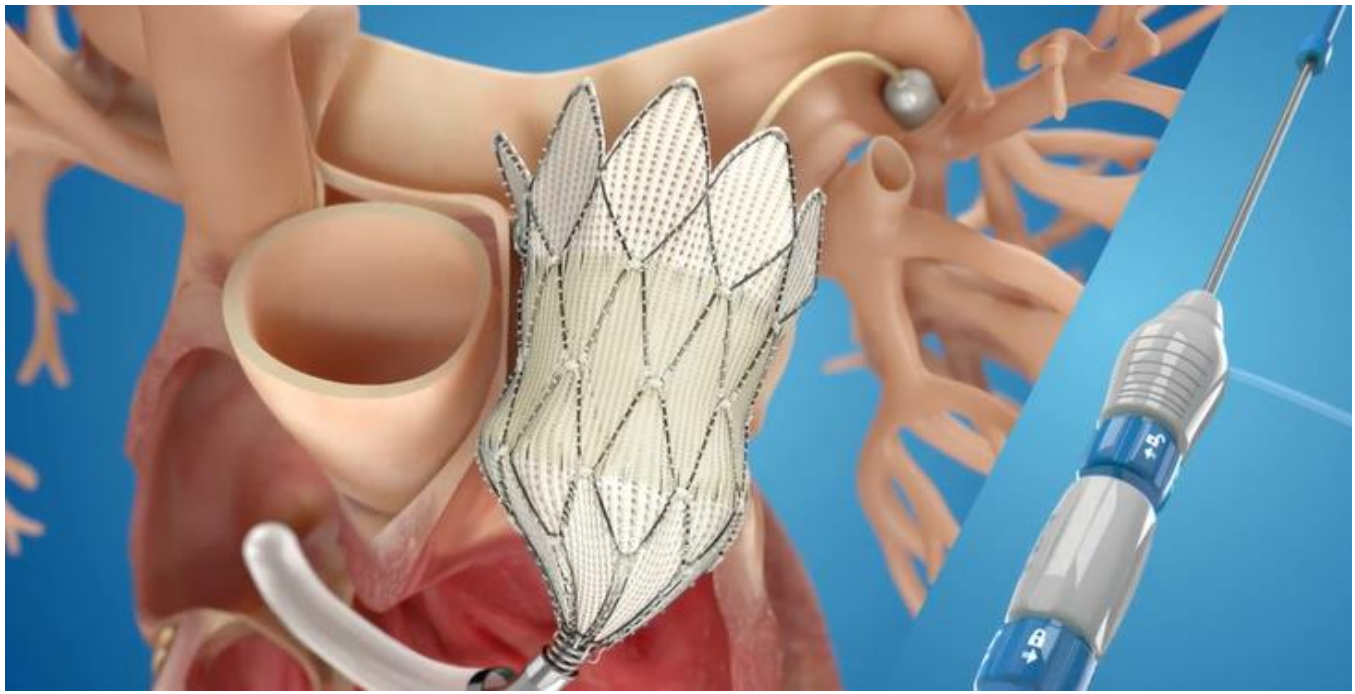




J Thorac Cardiovasc Surg 2011;142:1122-9.

Transcatheter pulmonary valve insertion (TPVI)

- Mess invasive
  - Not fit for all patient
  - Recurrence of PI/RVOTO
  - Ventricular arrhythmia



From Medtronic website

Conclusion

- The surgical repair for tetralogy of Fallot has an excellent outcome.
- The decision for palliation, the best palliation technique, the optimal timing for repair, and the best surgical repair technique, are still controversial.
- The long-term follow-up will be necessary to enhance the patient quality of life.

Thank you