22. GÖĞÜS KALP DAMAR ANESTEZİ VE YOĞUN BAKIM DERNEĞİ ULUSAL KONGRESİ

Tek ventrikül patofizyolojisi cerrahisi ve anestezi yönetimi

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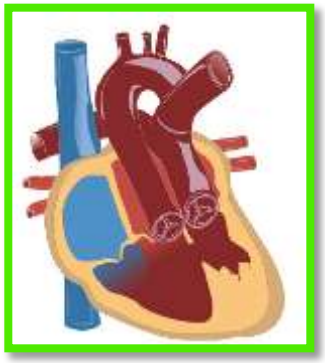
Anestezi ve Reanimasyon AD
İstanbul

Tanım

- Biventriküler düzeltmenin yapılamadığı kalp defektlerinde tek ventrikül fizyolojisi uygulanabilir

Tek ventrikül %

- Tek ventrikül insidensi her 100 000 doğumda 4-8 arasında ⋆
- Tüm konjenital kalp hastalıklarının %7.7 ⋆
  - %3.2 Hipoplastik sol kalp sendromu
  - %1.3 Trikuspit atrezisi
  - %3.2 Tüm diğer patolojiler

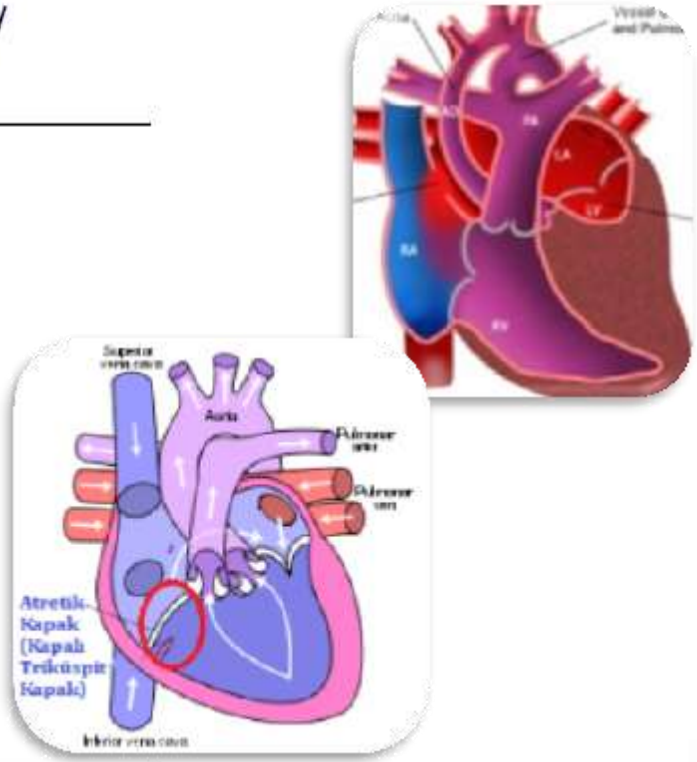


⋆ O'Leary PW . Prevalence, clinical presentation and natural history of patients with single ventricle. Progress in Pediatric Cardiology 16 (2002) 31–38

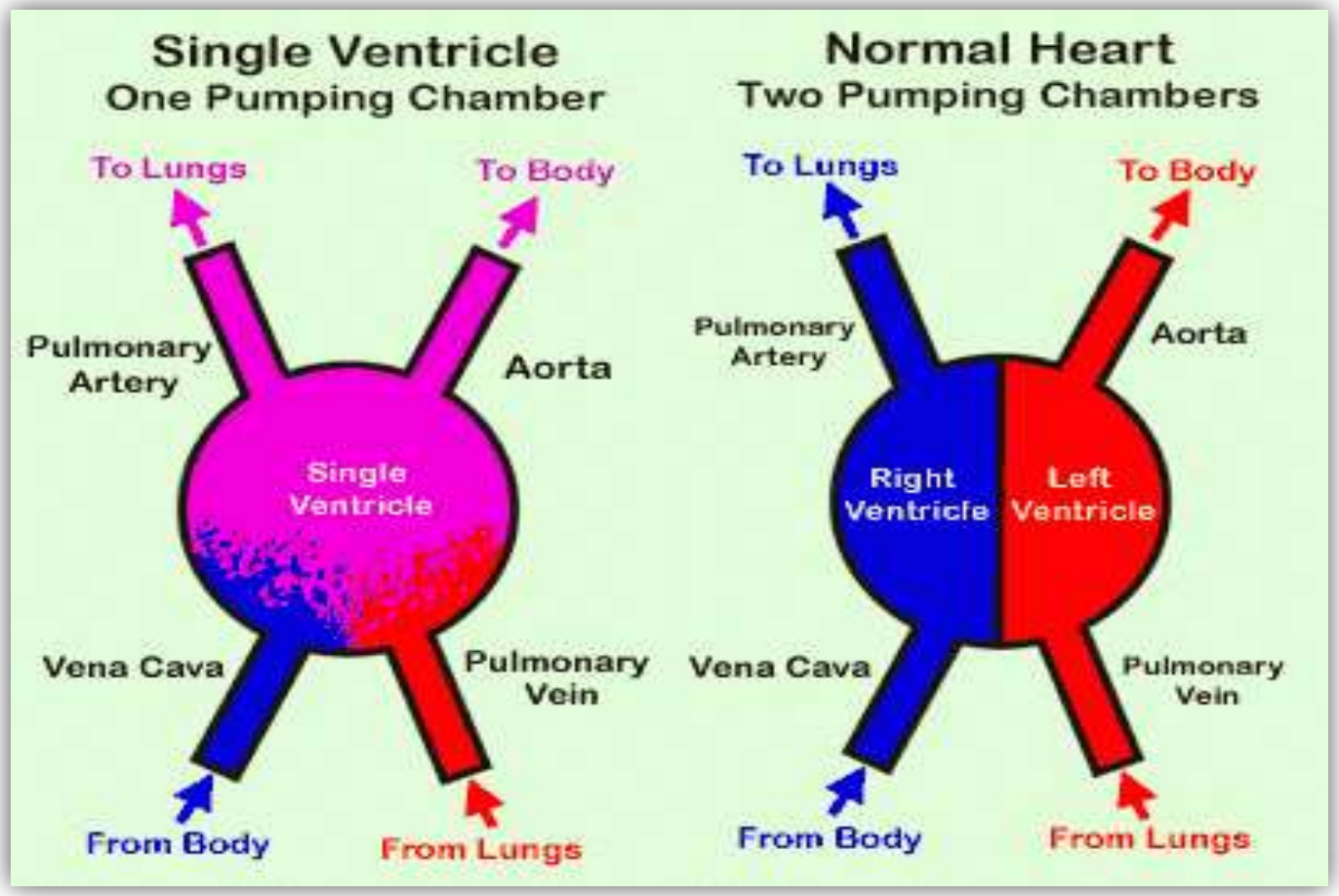
Tek ventrikül Anatomisi

Table 1. Diagnostic Terms for Patients With Functionally Univentricular Hearts.[a] *conj anomali*

1. Single ventricle, DILV
2. Single ventricle, DIRV
3. Single ventricle, Mitral atresia
4. Single ventricle, Tricuspid atresia
5. Single ventricle, Unbalanced AV canal
6. Single ventricle, Heterotaxia syndrome
7. Single ventricle, Other
8. Single ventricle + TAPVC
9. HLHS
10. Aortic valve atresia



Tek ventrikül fizyolojisi



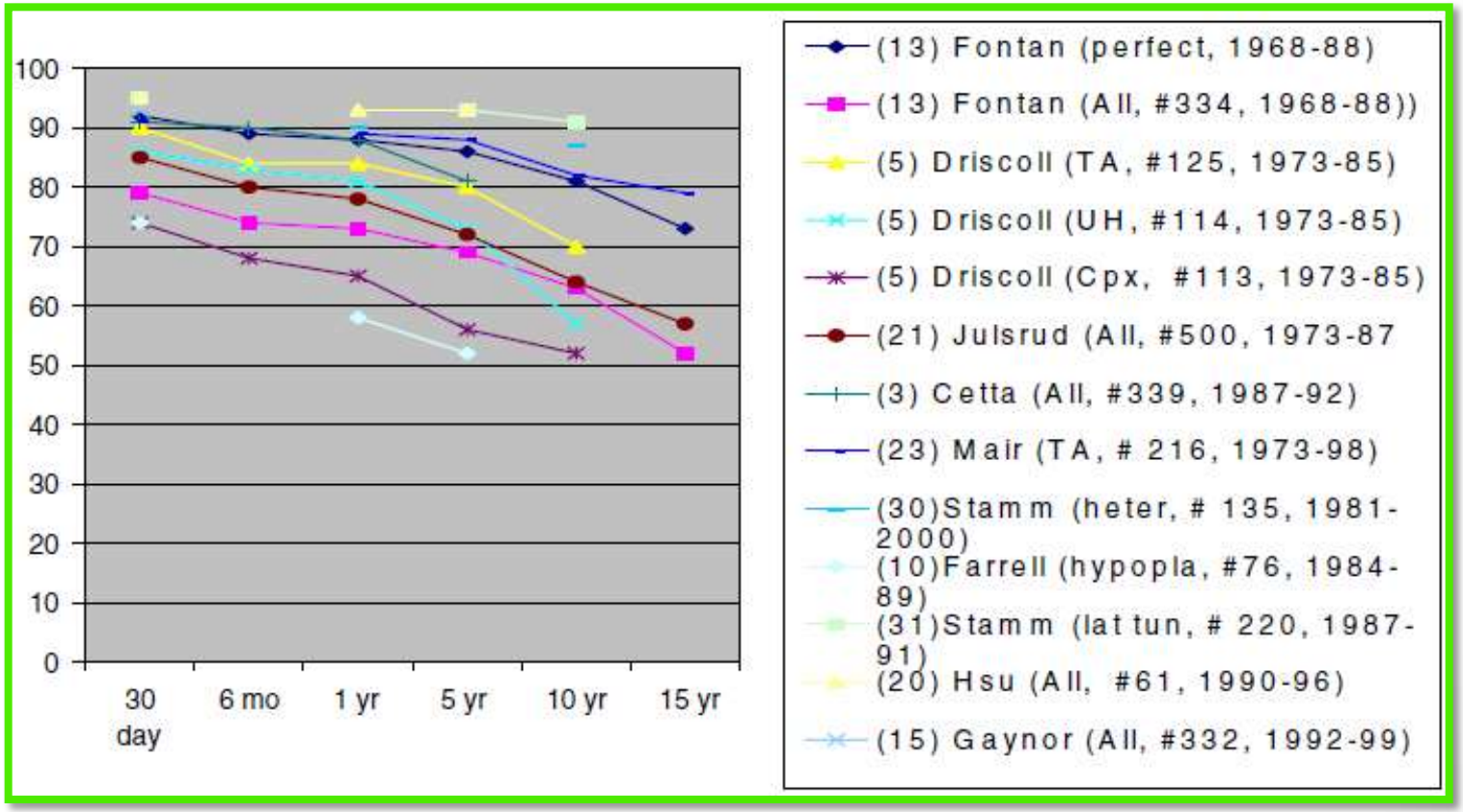
Opere edilmemiş tek ventrikül olgularında sağ kalım

Moodie ve ark* 83 olgu en geniş seri

- Hipoplastik sol ventrikül morfolojili
  - 4 yılda sadece %50'si hayatta kalır
- Hipoplastik sağ ventrikül morfolojili
  - %70'i 16 yaşından önce kaybedilir
- En önemli ölüm sebepleri
  - Aritmi, KKY, ani açıklanamayan ölüm

*Moodie DS et al. Long-term follow-up in the unoperated univentricular heart. Am J Cardiol. 1984;53:1124

Fontan ameliyatından sonra sağ kalım



Predictors of Survival After Single-Ventricle Palliation

The Impact of Right Ventricular Dominance

Yves d'Udekem, MD, PhD,* Mary Y. Xu, BMedSci,* John C. Galati, BSc, PhD,† Siming Lu, BMedSci,* Ajay J. Iyengar, MBBS, BMedSci,* Igor E. Konstantinov, MD, PhD,* Gavin R. Wheaton, MD,‖ James M. Ramsay, MD,¶ Leeanne E. Grigg, MBBS,# Johnny Millar, MB, ChB, PhD,‡ Michael M. Cheung, MD,§ Christian P. Brizard, MD*

(J Am Coll Cardiol 2012;59:1178–85)

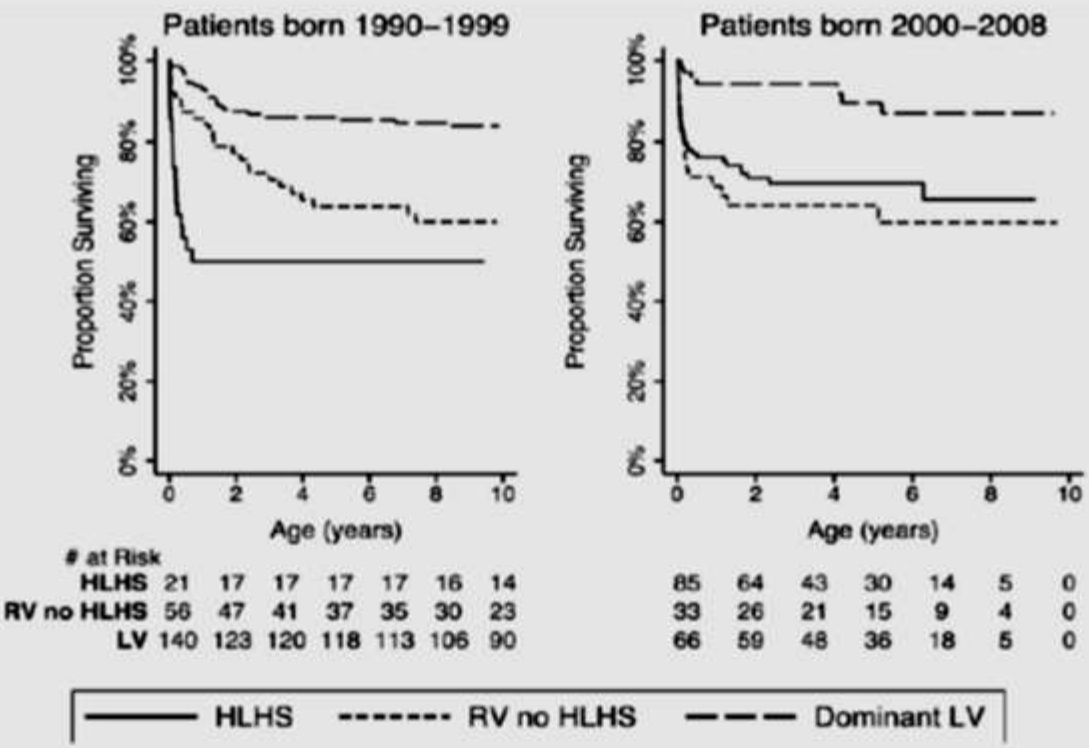


Figure 2 **Kaplan-Meier Survival Curves**

Patients were separated into 2 groups according to era of birth. Era 1 consisted of patients born from 1990 to 1999; era 2 consisted of patients born from 2000 to 2008. HLHS = patients with hypoplastic left heart syndrome; LV = patients with a dominant left ventricle; RV no HLHS = patients with a dominant right ventricle but not a diagnosis of HLHS.

1950--- ----------2016

- *Cerrahi aşamalar*
  - *Anestezi uygulamaları*

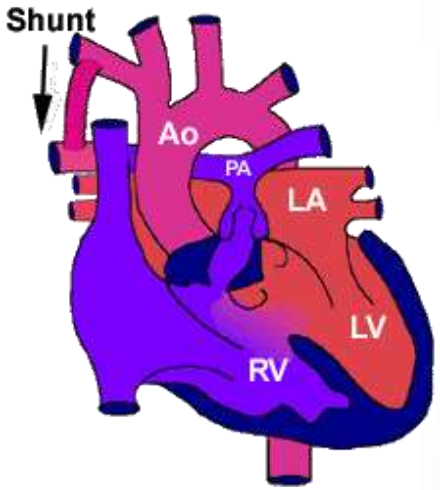
İlk aşama: Yenidoğan→ Qp/Qs

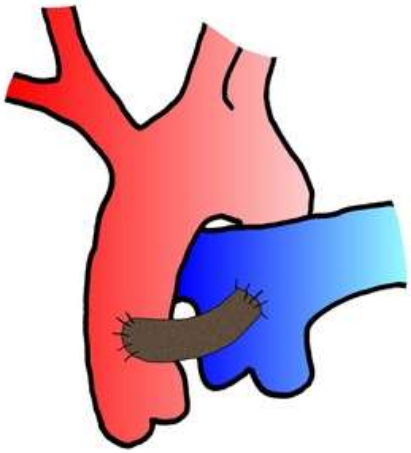
İkinci: BDG veya hemifontan: 4-6 ay paralel→ kısmi seri

Üçüncü:Fontan:15-24 aydan sonra: sistemik- kavapulmoner ayrılır

İlk aşama: Yenidoğan palyasyonu Pulmoner kan akımı yetersiz

- Qp düşük: Siyanoz
- Pulmoner kan akımını arttırmak için
  - Şant ameliyatı
    - MBT şant, santral şant, sano şant
- Bazı ekipler duktus arteriosus stent



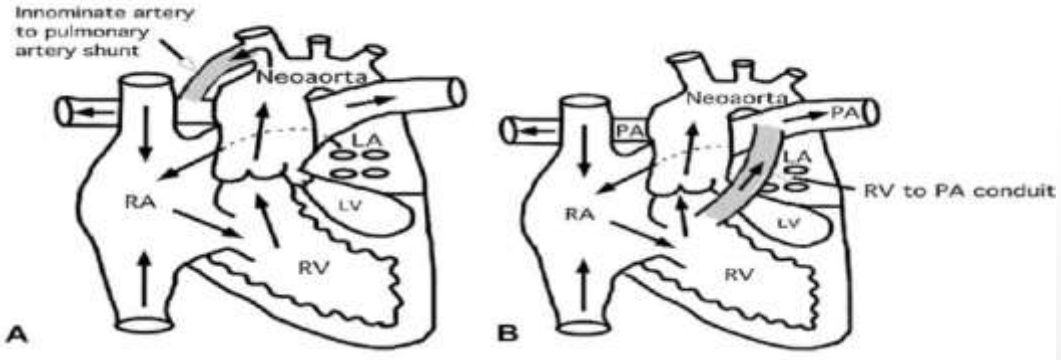


İlk Aşama: Yenidoğan palyasyon Sistemik kan akımı yetersiz

- Duktusa bağımlı sistemik dolaşım
  - PG infüzyonu
  - HLHS
    - NORWOOD
- Duktus kapanırsa
  - Sistemik hipoperfüzyon
    - Doku iskemisi, asidoz, NEK, BY, MOF, ex

Başarılı Norwood operasyonu

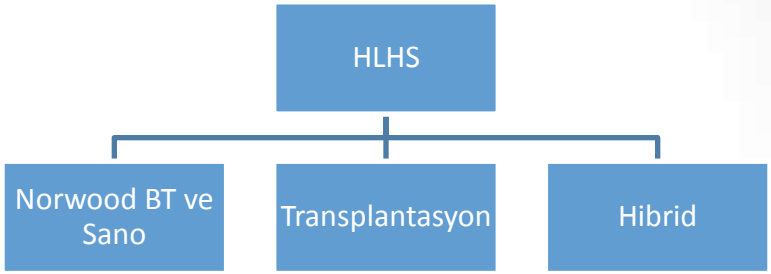
- Sorunsuz sistemik çıkış yolu
- Yeterli pulmoner kan akımı
  - MBT veya Sano şant
- Yeterli atrial karışım
  - Atrial septektomi
- Yeterli koroner perfüzyon



İlk Aşama: Yenidoğan palyasyonu

HLHS

- Norwood
  - Mortalite %50-%10-15 ↓
- Hibrid girişim
  - Mortalite % 10 kadar ↓
  - 1.2. aşamalar ilerleyen yaşta
- Tranplantasyon
  - Palyasyonun başarısız



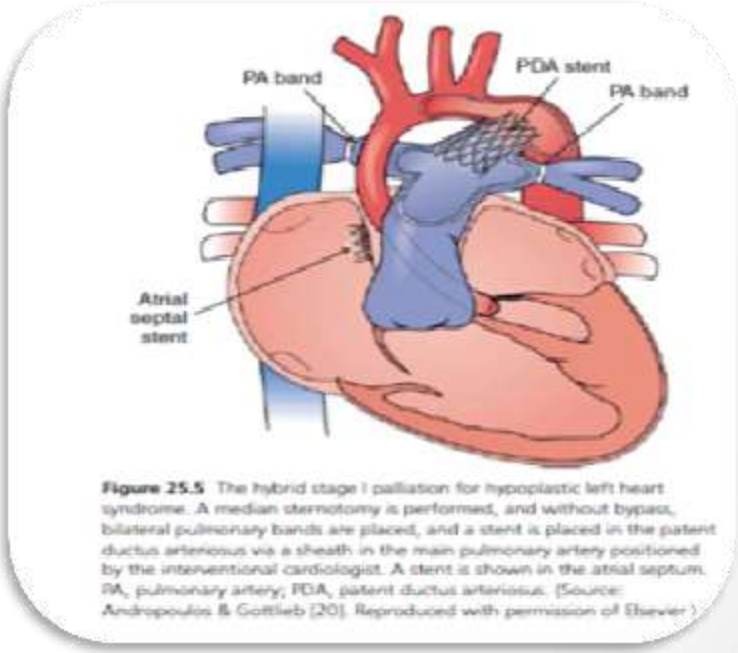
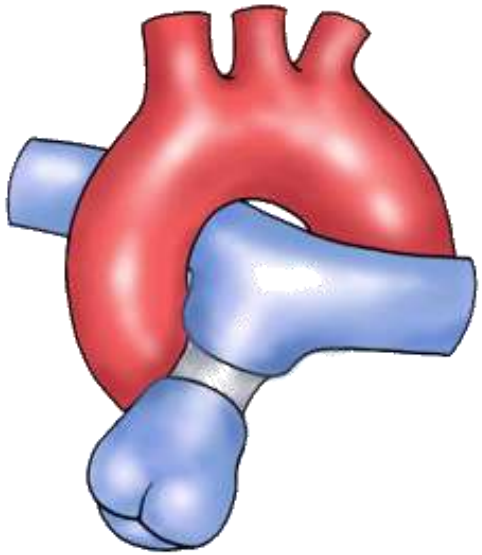


Figure 25.5 The hybrid stage I palliation for hypoplastic left heart syndrome. A median sternotomy is performed, and without bypass, bilateral pulmonary bands are placed, and a stent is placed in the patent ductus arteriosus via a sheath in the main pulmonary artery positioned by the interventional cardiologist. A stent is shown in the atrial septum. PA, pulmonary artery; PDA, patent ductus arteriosus. (Source: Andropoulos & Gottlieb [20]. Reproduced with permission of Elsevier.)

*Jacobs ML, et al. J Thorac Cardiovasc Surg 1998;

İlk Aşama: Pulmoner kan akımı fazla

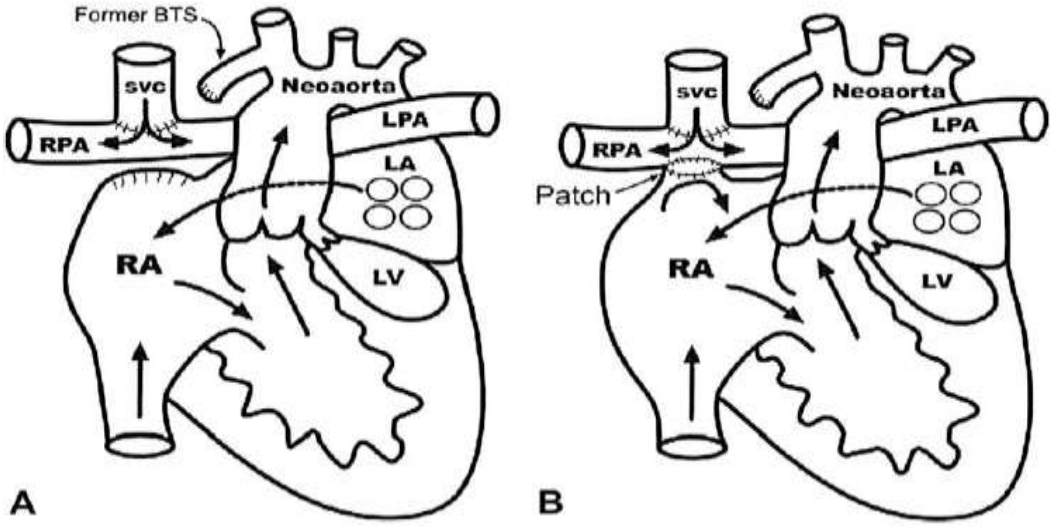
- Pulmoner kan akımı fazla
  - PHT (PVR yüksekse)
  - KKY (PVR düşükse)
- PAB



İkinci aşama: Süperiyor Kava-pulmoner anastomoz

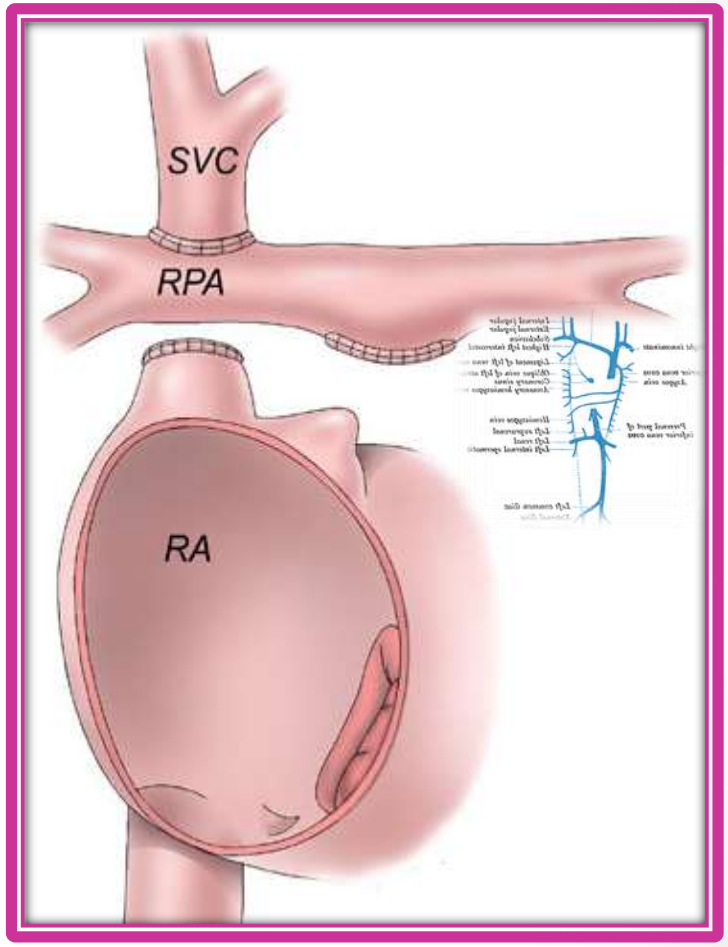
- Cerrahi: SVK- RA kesilerek PA lere anastomoz edilir.

  BDG veya Hemifontan



Bidirectional Glenn

- Kava-pulmoner anastomoz
  - Sadece İVK sistemik sirkülasyon
  - Volüm yükü azalır
  - Satürasyon: % 80-85
- Ameliyat 6.ayda
    - 2. aydan sonra (desatürasyon)
      - Pulmoner çap ve rezistans
- Çocuk büyüdükçe satürasyon ↓
  - İVK'den gelen kan artar
  - Venö-venöz kollateral
  - pulmoner arter- venöz sant
    - %25 olgu



Bidirectional Glenn: Preoperatif hazırlık

- **Pulmoner dolaşımda kanın akışına engel anatomik ve fonksiyonel yapı**
  - PVR ↓
  - AV valve fonksiyonu iyi
  - Ventriküler kompliyans yeterli
  - Atriyumlar arası yeterli geçiş
  - Yeterli PA indeksi
- **Aspirin kullanımı: postop kanama hazırlığı**

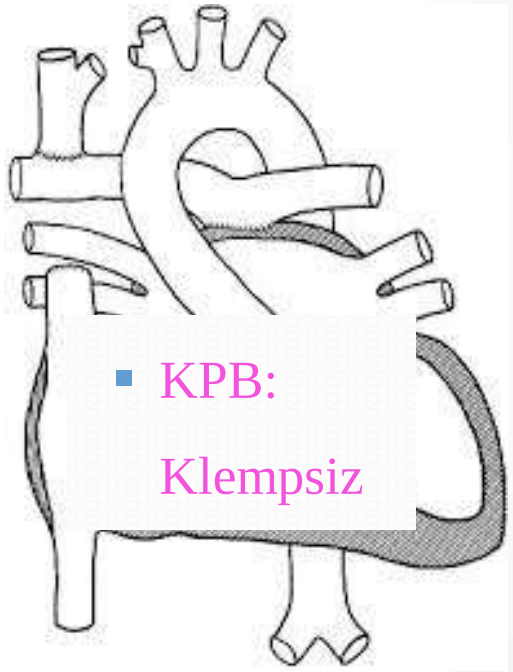
✓ **Pulmoner basınç hesaplanmalıdır.**

$$P_{SVC} = \left(\frac{(P_{PA} - P_{PV})(Q_{PA} : Q_{SA})}{Q_{PB} : Q_{SB}} \right) + P_{LA}$$

- Genel olarak ameliyat öncesi anjiografi ile PA P ölçülür
- Pulmoner arter basıncı: Max:15-16 kadar BDKPA yapılabilir

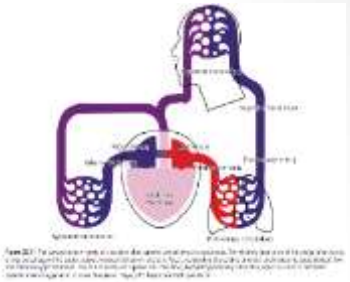
Bidirectional Glenn: İntraoperatif

- Premedik, İndüksiyon ve idame
  - Miyokard fonksiyonu, hemodinami
    - Çıkış yolu veya kapak fonk bzk
      - Normal dozda kısa etkili anestezik
      - Max:Fentanyl 5–10 µg/kg
    - Erken Ekstübasyon: SVK venöz dönüş
- İnvaziv monitörizasyon
  - Arter kanülü ve Venöz kateter yeri
    - Femoral ven ve 3 Fr. kateter SIJV
      - Anastamozun olmadığı taraf İJV
- KPB bitimi MOF
  - Kan kaybı, plevral ve perikadiyal efüzyonu azalttığı bildirilmektedir.



- KPB: Klempsiz

BDG: Postoperatif monitörizasyon



- Pompa Çıkışı: Yakın basınç monitörizasyon
- *Trans torasik sağ atrial kateter ve SVK erken çıkarılmalı*

Table 25.4 Hemodynamic status and differential diagnosis in the postoperative superior cavopulmonary anastomosis patient

Status	PAP(mmHg)	LAP(mmHg)	TPG(mmHg)	SpO_2 (%)	Etiologies
Normal	10–15	2–6 ↓	↓<10	80 ± 5	Ideal status
Elevated PAP	>15	2–6	>10	<75	High PVR; PA or PV obstruction
Elevated LAP	>15	>8–10	<10	<75	Ventricular systolic/diastolic dysfunction; sub-AS; AVVR; tamponade
Cyanosis	10–15	2–6	<10	<75	Decreased CBF; PV desaturation; decompressing veins; hypovolemia; anemia

NIRS

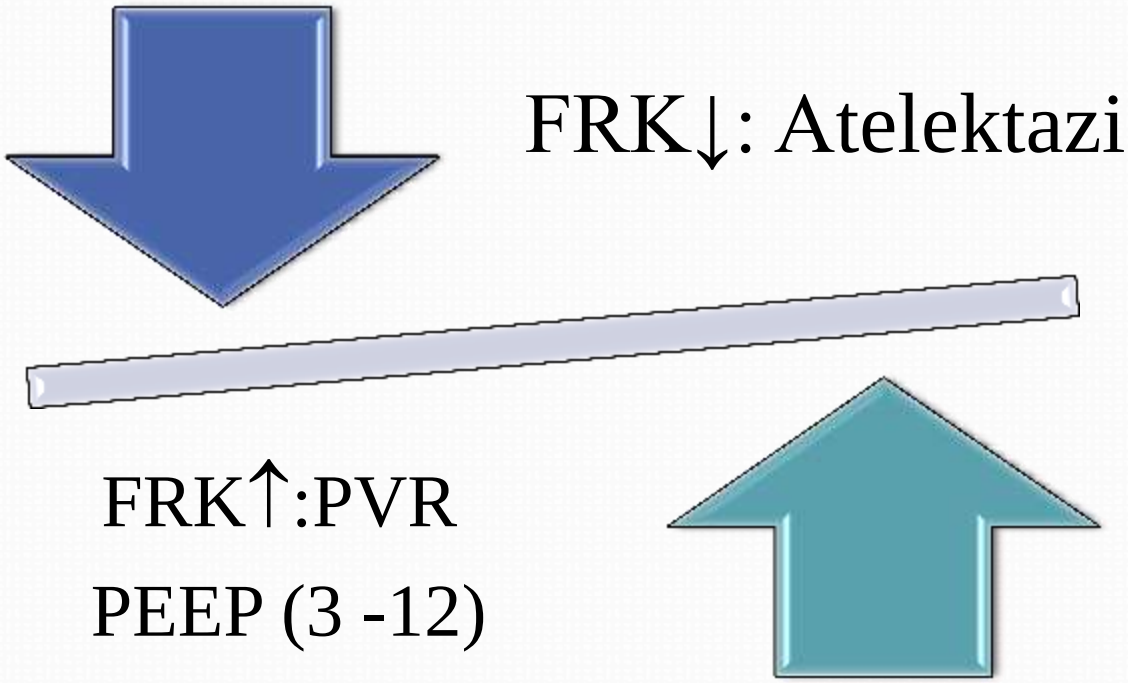
PAP, pulmonary artery pressure as measured in the superior vena cava; LAP, left or common atrial pressure; TPG; transpulmonary pressure gradient (PAP–LAP); PVR, pulmonary vascular resistance; PV, pulmonary vein; AS, aortic stenosis; AVVR, atrioventricular valve regurgitation; CBF, cerebral blood flow.

BDG:postoperatif Komplikasyonlar

- **Hipoksemi en sık (%70↓)**
  - **Hipovolemi** ve PBF azalması
  - Mekanik ventilasyon stratejisi
  - Venö-venöz kollateral ve AV malformasyon
- **Kavapulmoner dolaşım** P ↑
  - **SVKS**, siyanoz, CO azalma
  - $PaCO_2$ 40-45 mmHg:
    - **Serebral Venöz dönüş↑**
    - Erken ekstübasyon ve 30–45° baş nötr ve yukarı pozisyon
  - Medikal Tedavi
    - NO, sildenafil ilave edilir
  - Tekrar anjiyo p>18
- **Hipertansiyon ve bradikardi**
  - Beyin ↑venöz basınç ile konj
    - 4–8mmHg den 12–18mmHg
  - CPP azalmasına yanıt
    - Kan basıncının artar
  - Ani kalbe gelen kanın azalması
- **Düşük kardiak debi**
  - Aortik ark.obst, AV kapak Regürjitasyonu
  - Koroner perfüzyon ↓ veya miyokardiyal korunma yetersiz
  - Med ted. yanıt yok Transplantasyon

BDG: Postoperatif Ventilatör stratejisi

- Strateji: Dramatik PVR değişiklikleri **FRK korunarak yönetilmeli: SS, Pik P, İnsp süresi ↓**



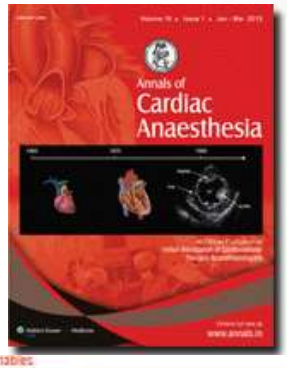
Original Article

This article is accompanied by an invited commentary by Prof. Muralidhar Kanchi

The effects of different ventilator modes on cerebral tissue oxygen saturation in patients with bidirectional superior cavopulmonary connection

Ayda Türköz, Şule Turgut Balcı, Hülya Gönen, Özlem Çınar, Emre Özker[1], Rıza Türköz[1]

Departments of Anesthesiology, and [1]Cardiovascular Surgery, Baskent University Istanbul Teaching and Medical Research Center, İstanbul, Turkey



ABSTRACT

Aims and Objectives: We used near-infrared spectroscopy to document changes in cerebral tissue oxygen saturation ($SctO_2$) in response to ventilation mode alterations after bidirectional Glenn (BDG; superior cavopulmonary connection) procedure. We also determined whether spontaneous ventilation have a beneficial effect on hemodynamic status, lactate and $SctO_2$ when compared with other ventilation modes. **Materials and Methods:** 20 consecutive patients undergoing BDG were included. We measured $SctO_2$ during three ventilator modes (intermittent positive-pressure ventilation [IPPV]; synchronized intermittent mandatory ventilation [SIMV]; and continuous positive airway pressure + pressure support ventilation [CPAP + PSV]). We, also, measured mean airway pressure (AWP), arterial blood gases, lactate and systolic arterial pressures (SAP). **Results:** There was no change in $SctO_2$ in IPPV and SIMV modes; the $SctO_2$ measured during CPAP + PSV and after extubation increased significantly (60.5 ± 11, 61 ± 10, 65 ± 10, 66 ± 11 respectively) ($P < 0.05$). The differences in the SAP measured during IPPV and SIMV modes was insignificant; the SAP increased significantly during CPAP + PSV mode and after extubation compared with IPPV and SIMV (109 ± 11, 110 ± 12, 95 ± 17, 99 ± 13 mmHg, respectively) ($P < 0.05$). Mean AWP did not change during IPPV and SIMV modes, mean AWP decreased significantly during CPAP + PSV mode (14 ± 4, 14 ± 3, 10 ± 1 mmHg, respectively) ($P < 0.01$). **Conclusions:** The $SctO_2$ was higher during CPAP + PSV ventilation and after extubation compared to IPPV and SIMV modes of ventilation. The mean AWP was lower during CPAP + PSV ventilation compared to IPPV and SIMV modes of ventilation.

Table 2: Summary of clinical variables

	Mean ± SD				P		
	IPPV	SIMV	CPAP+PSV	Extubation	IPPV-SIMV	IPPV-CPAP+PSV	IPPV-extubation
SpO_2	78 ± 10	77 ± 9	75 ± 8	77 ± 8	0.248	0.123	0.544
End-tidal CO_2	36 ± 5	36 ± 5	39 ± 5	-	0.186	0.147	-
$PaCO_2$	46 ± 4	46 ± 5	45 ± 5	46 ± 7	0.563	0.368	0.134
Hematocrit	37 ± 3	35 ± 2	36 ± 2	36 ± 3	0.068	0.093	0.098
SAP	95 ± 16	99 ± 14	109 ± 11	110 ± 12	0.703	0.039*	0.042*
DAP	50 ± 10	55 ± 8	60 ± 11	61 ± 10	0.057	0.009**	0.002**
MAP	67 ± 11	71 ± 11	79 ± 10	79 ± 10	0.138	0.007**	0.005**
Lactate	1.9 ± 1	2.2 ± 0.9	1.8 ± 0.6	2.5 ± 0.6	0.126	0.712	0.173
Mean airway pressure	14 ± 4	14 ± 3	10 ± 1	-	0.977	0.001**	-
Temperature	36 ± 0.6	36 ± 0.5	36 ± 0.6	36 ± 0.6	0.139	0.843	0.250
pH	7.3 ± 0	7.3 ± 0	7.3 ± 0	7.3 ± 0	0.117	0.063	0.067
CVP	15 ± 3	14 ± 3	13 ± 3	12 ± 2	0.078	0.003**	0.001**
Inotropic scores	9.6 ± 4	8.7 ± 5	7.3 ± 5	7.5 ± 6	0.147	0.052*	0.049*

*P < 0.05, **P < 0.01. IPPV: Intermittent positive-pressure ventilation, SIMV: Synchronized intermittent mandatory ventilation, SAP: Systolic arterial pressure, DAP: Diastolic arterial pressure, MAP: Means arterial pressure, CPAP: Continuous positive airway pressure, CVP: Central venous pressure, PSV: Pressure support ventilation, SD: Standard deviation

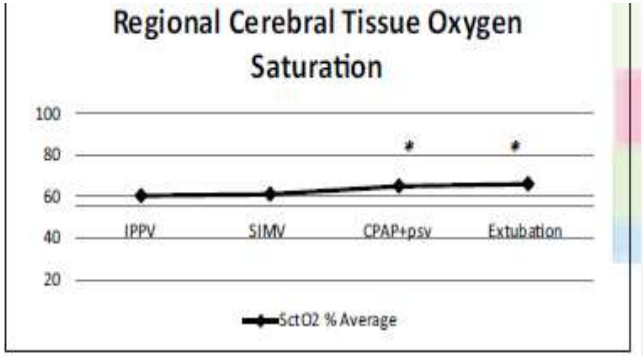


Figure 1: Changes in regional cerebral tissue oxygen saturation in intensive care unit *$P < 0.05$

Bidirectional Glenn: Postoperatif Hemodinami

- Hemodinamiyi olumlu etkiler
- Paralel sirkülasyonun yerini seri alır
- RV venöz dönüşü azalır
  - Volum output ↓
- Kalbin işi (Qs) %50↓
  - Dopamine (1–3 μg/kg/min)
- Diyastolik disfonksiyon

  İnodilator (milrinone)

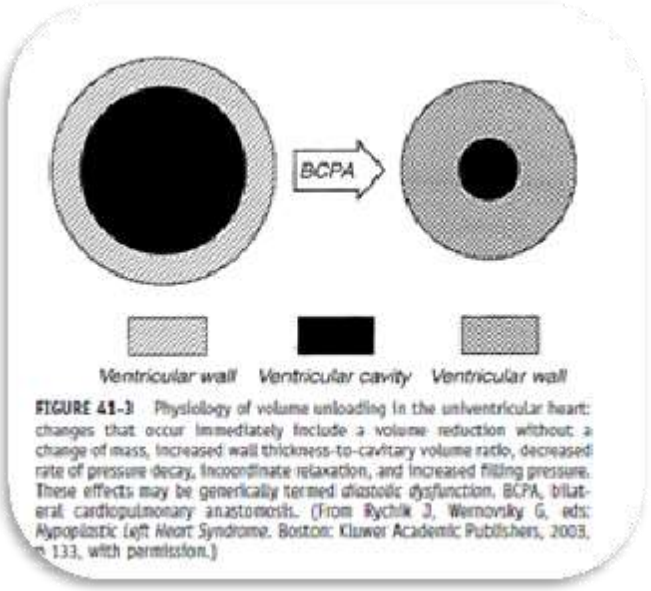


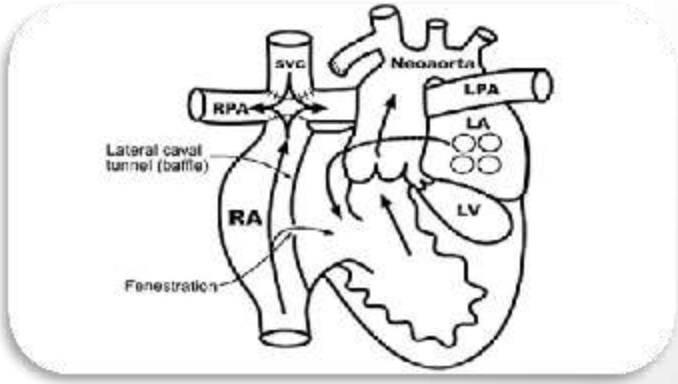
FIGURE 41-3 Physiology of volume unloading in the univentricular heart: changes that occur immediately include a volume reduction without a change of mass, increased wall thickness-to-cavitary volume ratio, decreased rate of pressure decay, incoordinate relaxation, and increased filling pressure. These effects may be generically termed *diastolic dysfunction*. BCPA, bilateral cardiopulmonary anastomosis. (From Rychik J, Wernovsky G, eds: *Hypoplastic Left Heart Syndrome*. Boston: Kluwer Academic Publishers, 2003, p 133, with permission.)

BDG:Sonuçlar

- Mortalite % 4
- Erken en (3 ay) SKPA yapılanlarda mortalite farkı yok
  - YB süresi↑
- SKPA sonrası fontan bekleme döneminde risk faktörleri
  - TV rejürjitasyonu, düşük ağırlık

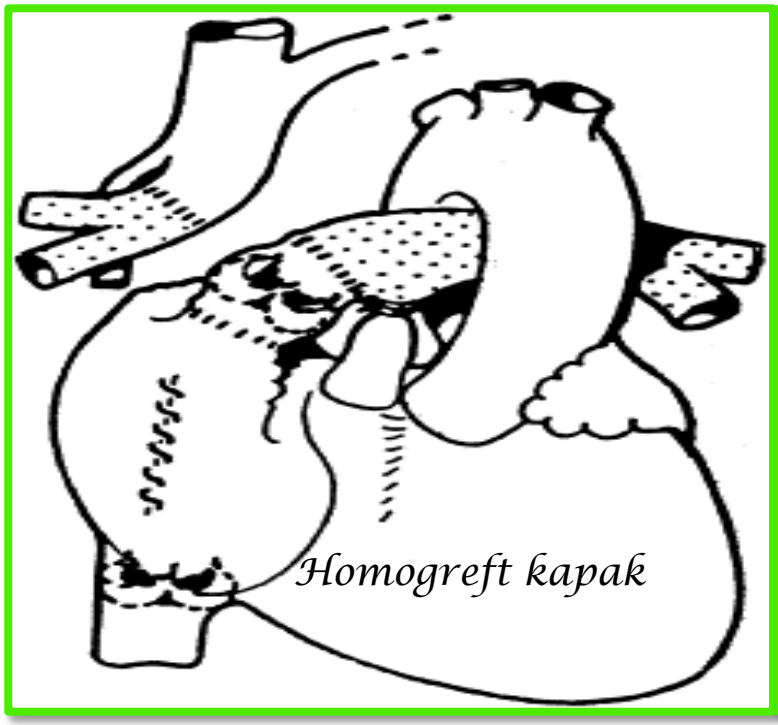
Fontan ameliyatı: Üçüncü evre

- Yaş > 18- 24 ay
- İVK ile pulmoner artere birleştirilir
- Total kava-pumoner anastomoz
  - ***İki yöntem: Lateral tünel & ektrakardiyak konduit***
  - ***PAP artışına önlem: Fenestrasyon***
    - **Greftten sağ atriuma geçiş**

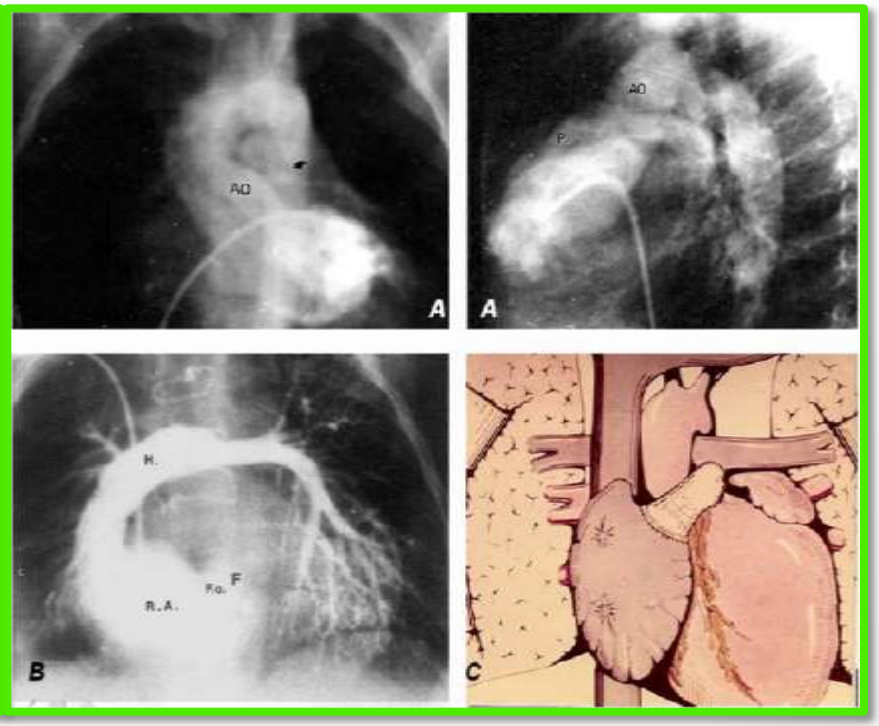


Fontan ameliyatı: Tarihçe

Orjinal FONTAN: 1971



KREUTZER Modifikas 1973-1982



Günümüzde Fontan varyasyonları

- **Lateral tünel veya Ektrakardiyak konduit**

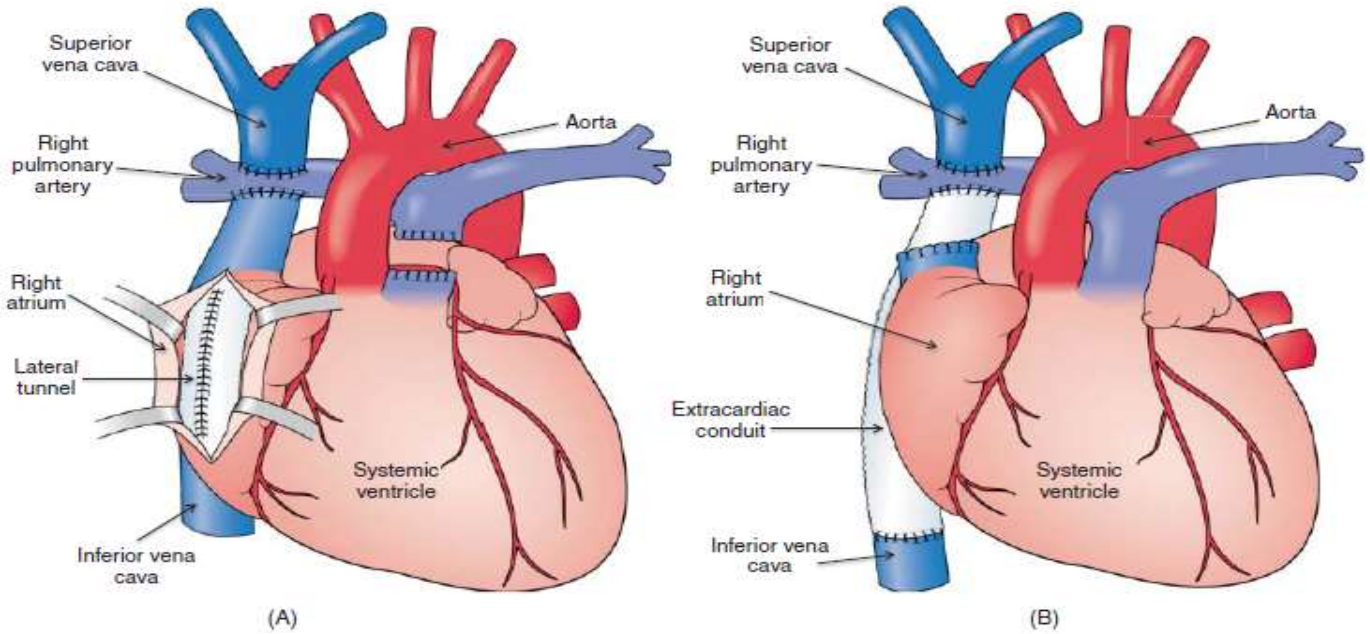
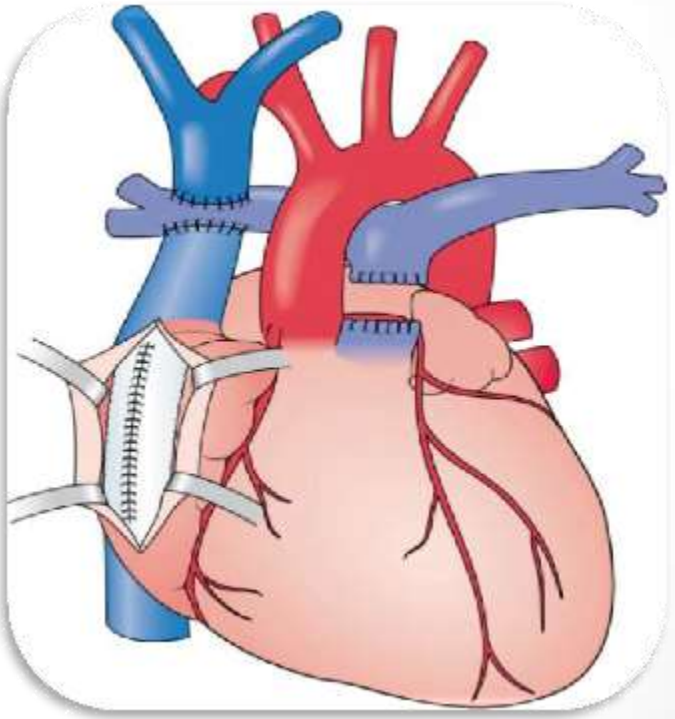


Figure 25.12 A. Lateral tunnel Fontan total cavopulmonary connection. (A) A polytetrafluoroethylene (PTFE) patch or tube is fashioned inside the right atrium. (B) Extracardiac conduit Fontan. The PTFE tube is anastomosed outside the heart from the detached inferior vena cava directly to the right pulmonary artery. (Source: Andropoulos et al. [20]. Reproduced with permission of Elsevier.)

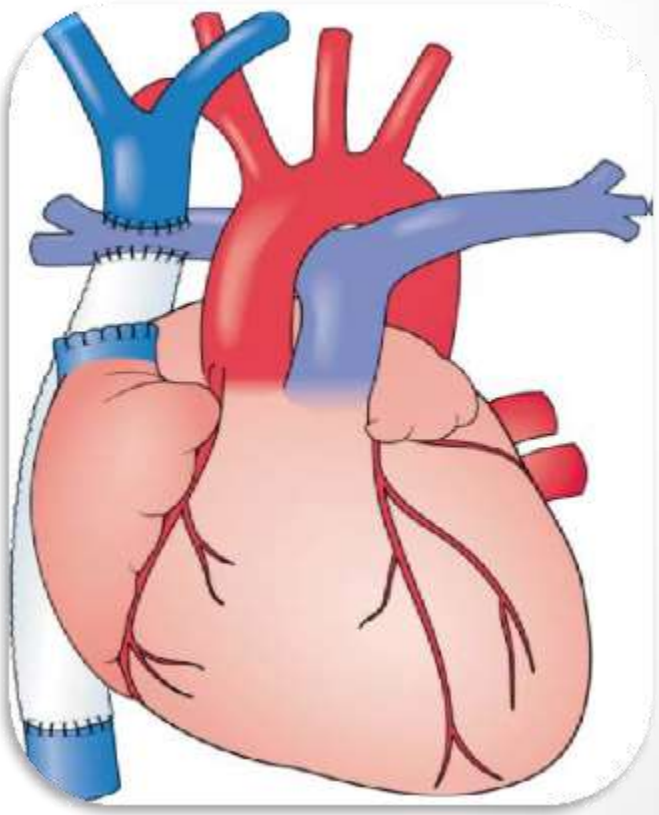
Fontan ameliyatı: Lateral tunel

- 1990-2000 yıllarında en sık intra-atrial PTFE tunnel
  - İVC ile SVC birleştirilir
  - Sağ kalp açılır
    - KPB ve klemp tekniği



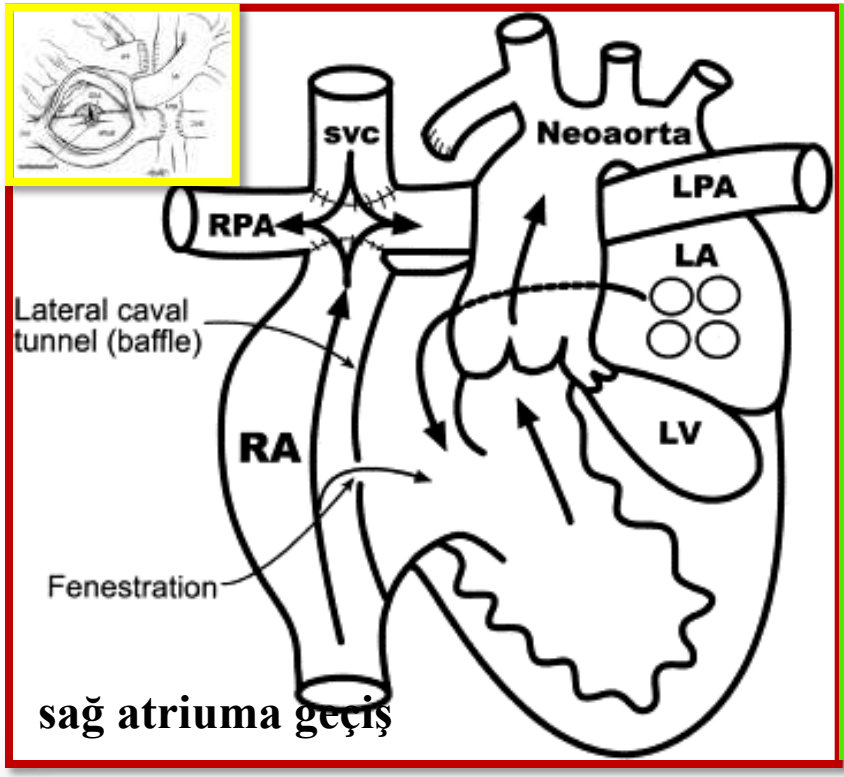
Fontan : Ektrakardiyak conduit

- 2000 den sonra sıklıkla Extrakardiyak fontan
- 18-20mm PTFE tüp greft kalbin dışından IVK ve sağ PA alt kısım ile anastomoz
  - KPB (Klempsiz)
  - KPBsız: sağ kalp bypassı ile kan transfüzyonu azaltılarak yapılabilir

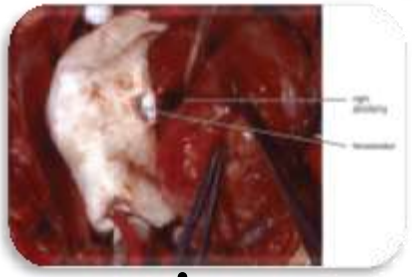


Fontan: Ekstrakardiak & Lateral Tunel Fenestrasyon

- *Boston grubu tarafından 1990, Kavapulmoner anastomozda basınç artışı :*
- *Sistemik venöz P azaltır, Preload ve CO sürdürür ve Sistemik Satürasyon düşer*



Fontan: Fenestrasyon ?



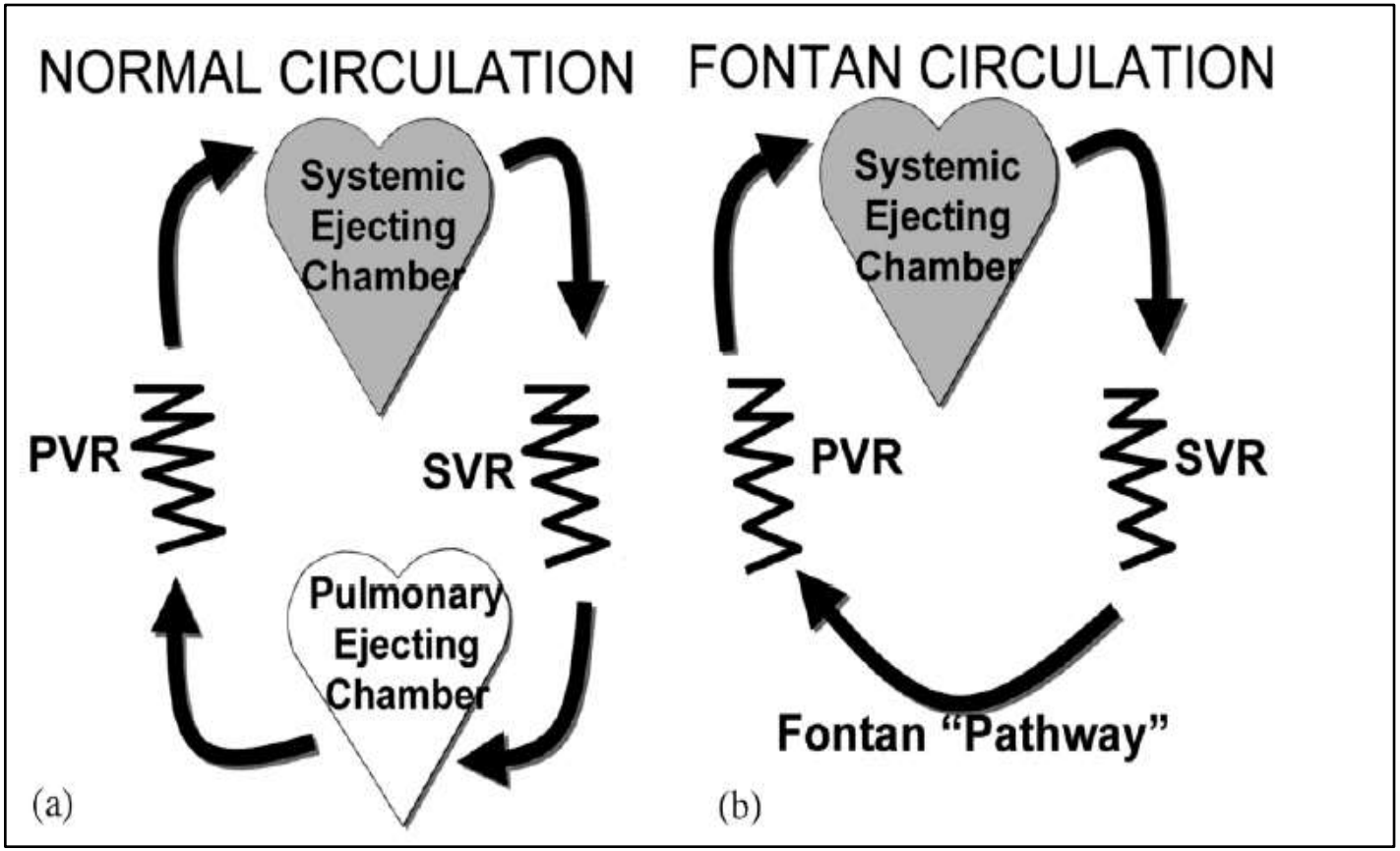
Avantaj

- Postop hemodinami iyi
- YB süresi ↓
- Toraks tüpü süresi ↓
- Taburcu süresi ↓
- Venöz sistemden anjio ve kateter

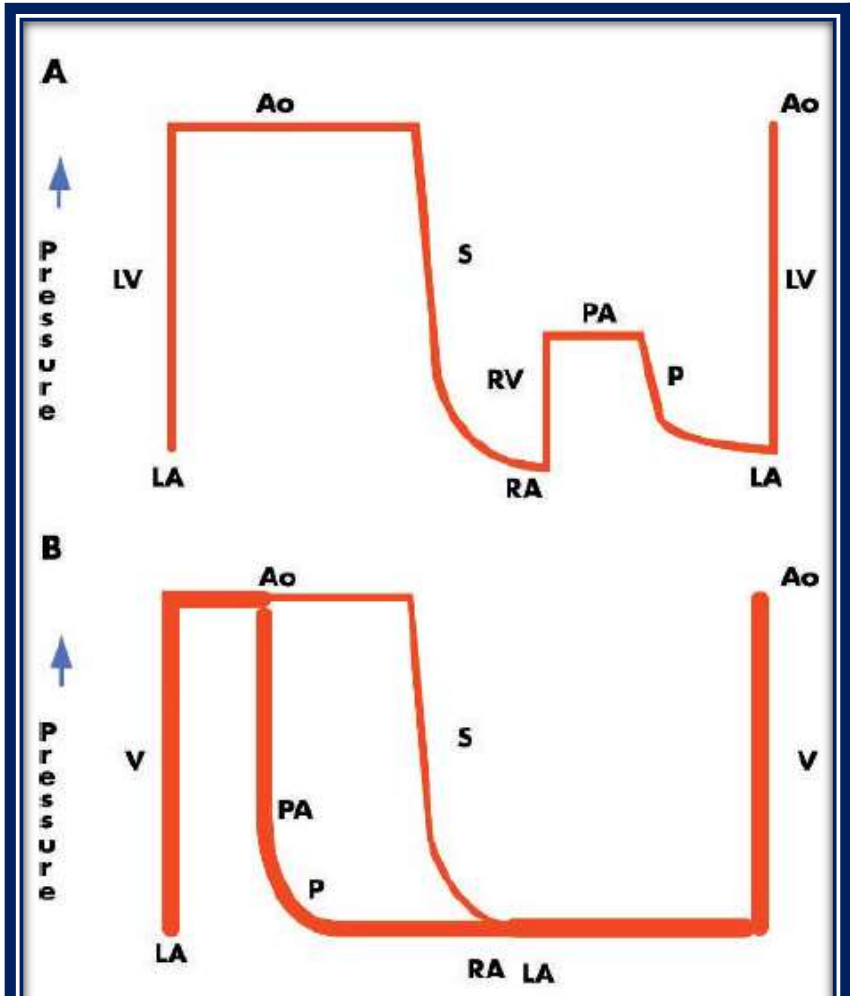
Dezavantaj

- Siyanoz
- Tromboemboli**
- Kapamak için girişim
- Aritmi ?
- Hipoksemi
- polisitemi

Fontan Fizyolojisi



Fontan Fizyolojisi



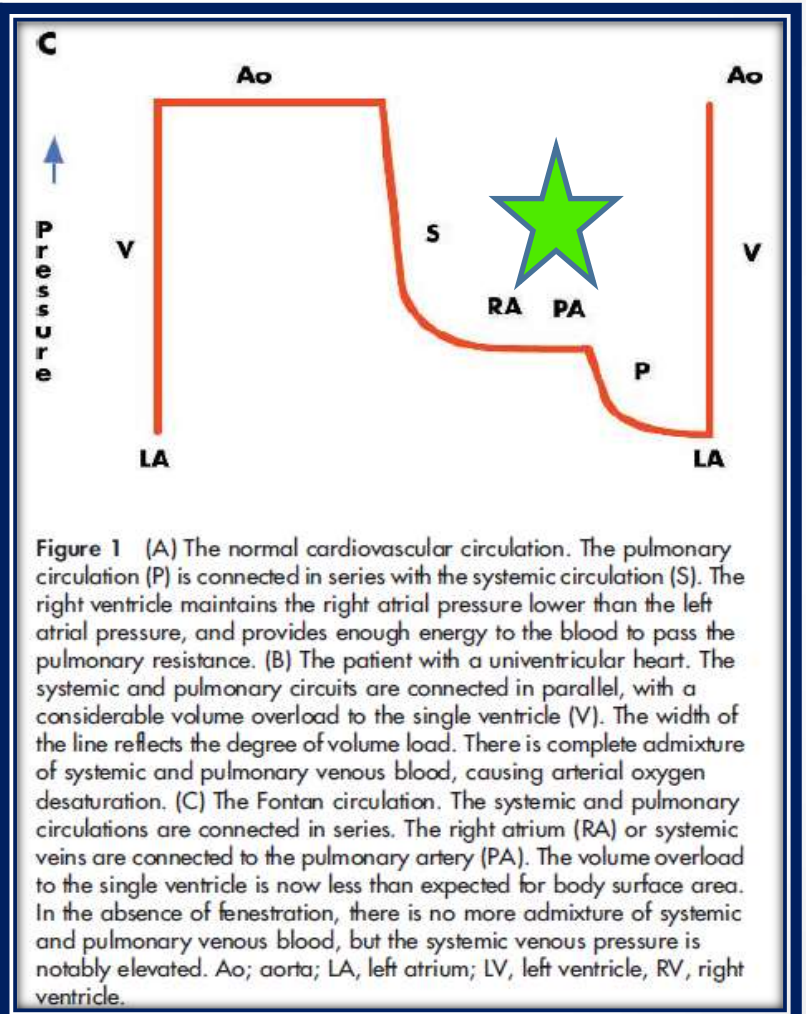


Figure 1 (A) The normal cardiovascular circulation. The pulmonary circulation (P) is connected in series with the systemic circulation (S). The right ventricle maintains the right atrial pressure lower than the left atrial pressure, and provides enough energy to the blood to pass the pulmonary resistance. (B) The patient with a univentricular heart. The systemic and pulmonary circuits are connected in parallel, with a considerable volume overload to the single ventricle (V). The width of the line reflects the degree of volume load. There is complete admixture of systemic and pulmonary venous blood, causing arterial oxygen desaturation. (C) The Fontan circulation. The systemic and pulmonary circulations are connected in series. The right atrium (RA) or systemic veins are connected to the pulmonary artery (PA). The volume overload to the single ventricle is now less than expected for body surface area. In the absence of fenestration, there is no more admixture of systemic and pulmonary venous blood, but the systemic venous pressure is notably elevated. Ao; aorta; LA, left atrium; LV, left ventricle, RV, right ventricle.

Fontane anestezi yönetimi

- Preop: **Pulmoner dolaşımda kanın akışına engel anatomik ve fonksiyonel yapı**

$$P_{SVC} = \left(\frac{(P_{PA} - P_{PV})(Q_{PA} : Q_{SA})}{Q_{PB} : Q_{SB}} \right) + P_{LA}$$

= 14-15 mmHg geçmemelidir

- Premedikasyon: Yaş kritik (midazolam)
- İndüksiyon ve idame: BDG ile aynı
- İnvaziv monitörizasyon:
  - Arter ve Venöz kateteri yeri: femoral ven ve 3 Fr kat IJV
- Erken Ekstübasyon: Normotermi, hemodinami ve kanama kontr.

Fontane Postoperatif monitörizasyon

- *Erken post-operatif tedaviside amaç mümkün olan en düşük santral venöz basınç ile kalp debisini normal değerlerde sürdürmektir*

Table 25.5 Hemodynamic status and differential diagnosis of abnormalities in the postoperative Fontan patient

Status	PAP (mmHg)	LAP (mmHg)	TPG (mmHg)	Systolic BP (mmHg)	SpO_2	Etiology
Normal	10–15	2–6	<10	85–95	95±5	Ideal status
Decreased PAP and LAP	8–10	0–4	<10	<80	90±5	Hypovolemia
Elevated PAP	>15	2–6	>10	80±5	90±5	High PVR; PA or PV obstruction
Elevated LAP	>15	>8–10	<10	80±5	90±5	Ventricular systolic/diastolic dysfunction; AV dissociation; AVVR; tamponade
Cyanosis	10–15	2–6	<10	85–95	<85%	Excessive fenestration size or Fontan baffle leak; PV desaturation; decompressing veins; hypovolemia; anemia

NIRS

Fontan Postoperatif yönetim

- Sistemik oksijenizasyon **sistemik venöz basınç artışı** ile sağlanır
  - Pozisyon, mobilizasyon ve enteral beslenme
  - Mekanik ventilasyon stratejisi 2. aşama ile aynı
    - Erken ektübasyon şart ancak engel durum varsa
    - PEEP: FRK normal değerlerinde tutmak için kullanılır
    - **Hiperventilasyon, atelektazi ve PVR engellenmeli**
    - Tedaviye yanıt vermeyen düşük satürasyon ve yüksek Pulmoner basınç: NO****vent fonk göre
- İnotrop kullanımı
  - Düşük doz inotrop ihtiyacı
- Antikoagulasyon: erken dönemde rijit protokol

Fontan sonrası komplikasyonlar

ERKEN KOMPLİKASYONLAR

- **Kava-pulmoner dolaşımda P artmasına (P:18**)**
  - SVCS, hepatomegali, asit, ödem, siyanoz (fens ile)
- **Düşük kardiak debi**
  - Med tedavi, assist device, takedown, transplantasyon
- **Hipoksemi %90 ↓**
- **Akut KC disfonksiyonu**
- **Aritmi**: tolere etmez. AV senkroni vitaldir.

- **Diğerleri**
  - Plevral effüzyon, şilotoraks, perikardial effüzyonu
  - Frenik sinir paralizisi

GEÇ Kronik KOMPLİKASYON

- Venöz kollateral gelişimi, intrahepatik venöz fistüller , PAVM, sistemik-pulm arterial kollateral
- Gelişme geriliği
- Tromboembolik olaylar
- **PLE: Prt kayb (Albümine ↓)**
- **Plastik bronşitis,**
  - proteinli cast

Nonkardiyak cerrahide fontan olguları

Table 1. Long-term morbidities of the Fontan procedure
Arrhythmia
Protein-losing enteropathy
Plastic bronchitis
Hepatic cirrhosis
Thrombosis and thromboembolism
Pulmonary hypertension
Heart failure

Fontan: Sonuçlar

- Mortalite %2 ↓
  - KPB süresi
  - VEDP ↑
  - AV kapak yetmezliği
  - Heterotaxy
  - 2. ve 3. aşamanın birlikte

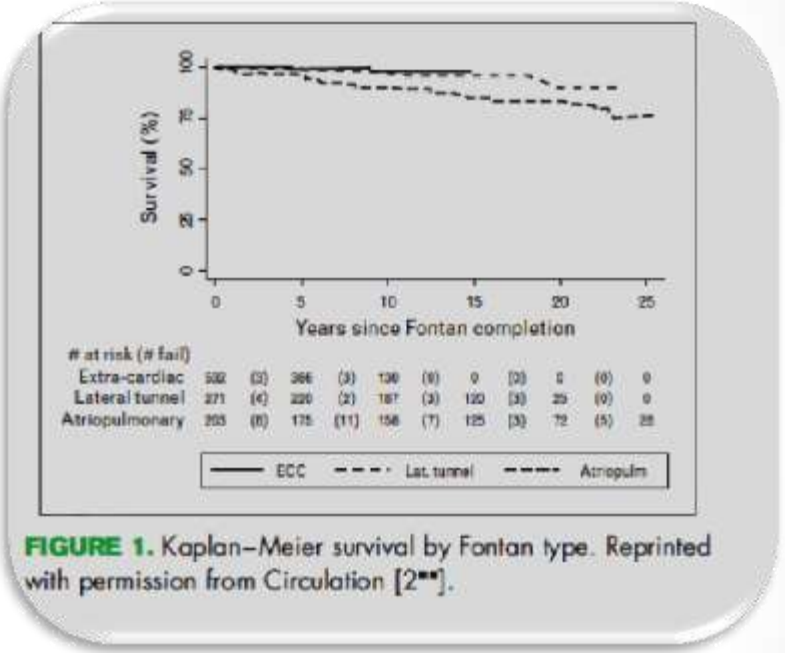
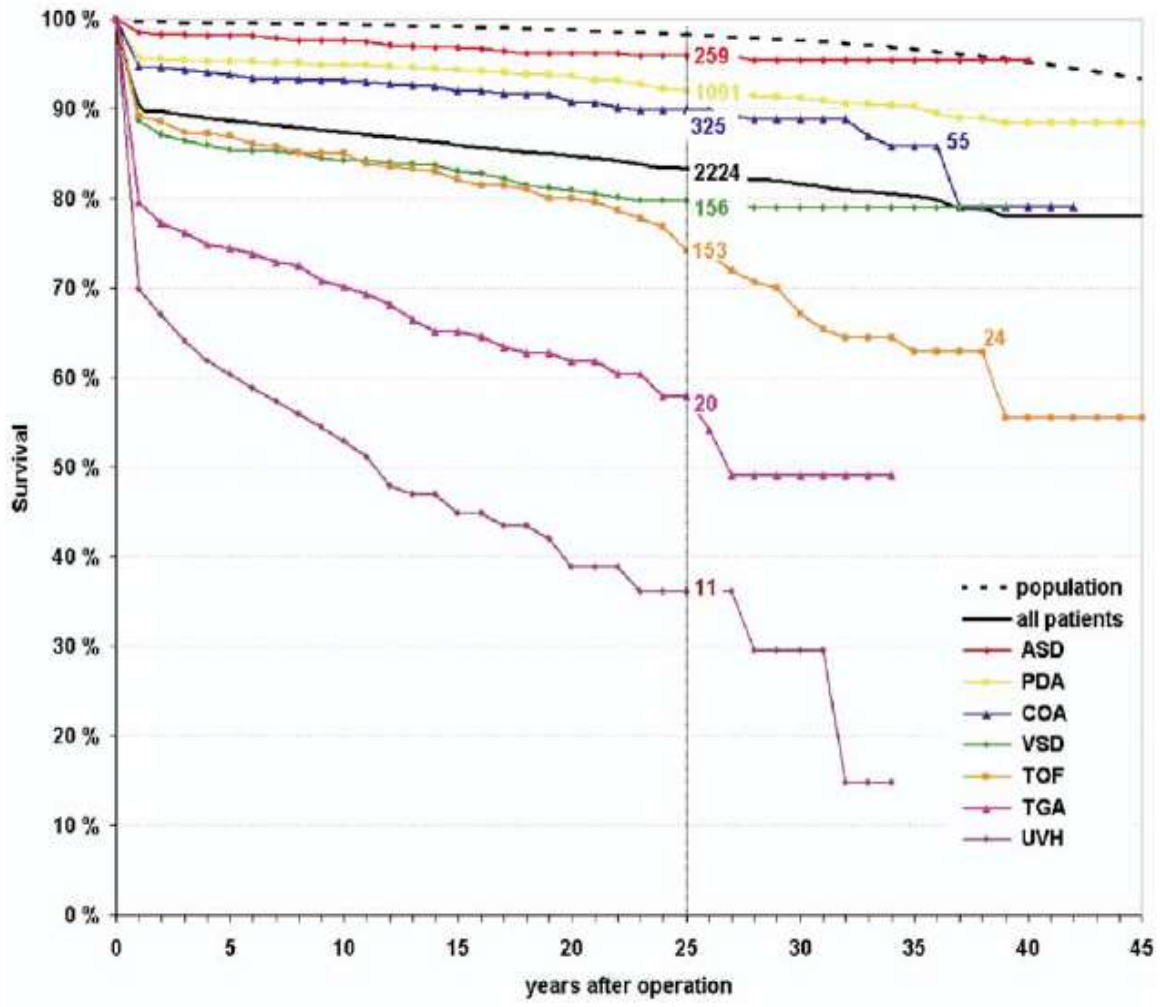


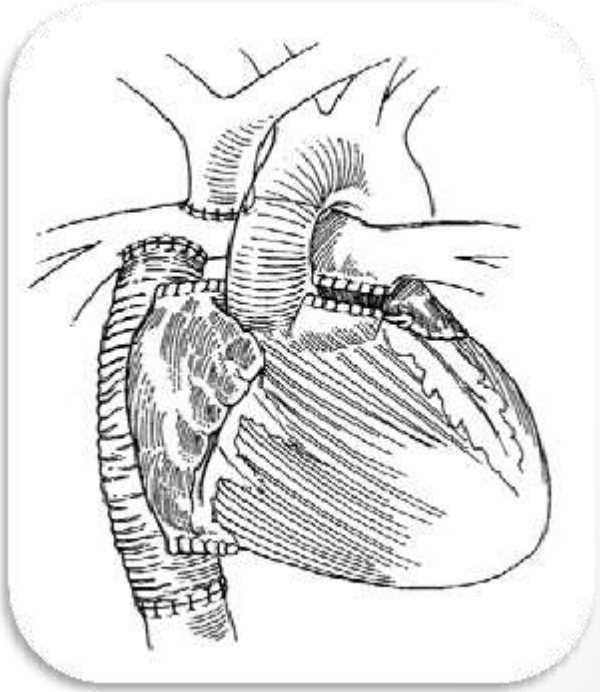
FIGURE 1. Kaplan–Meier survival by Fontan type. Reprinted with permission from Circulation [2▪▪].

Pediatric Cardiac Surgery Annual

Late Results and Quality of Life After Pediatric Cardiac Surgery in Finland: A Population-Based Study of 6,461 Patients With Follow-Up Extending up to 45 Years

Heikki I. Sairanen, Heta P. Nieminen, and Eero V. Jokinen





TEŞEKKÜR EDERİM

Damus-stansel-Kaye procedure

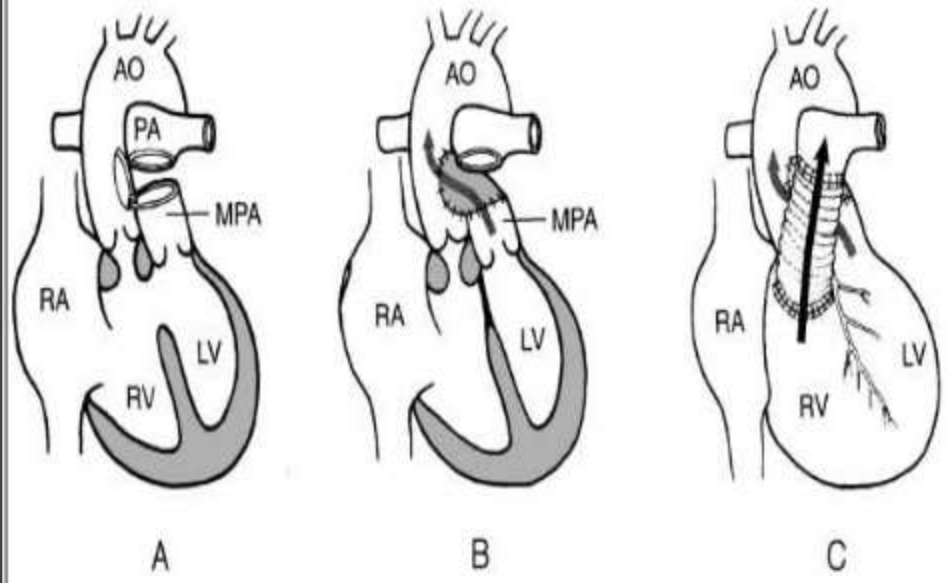
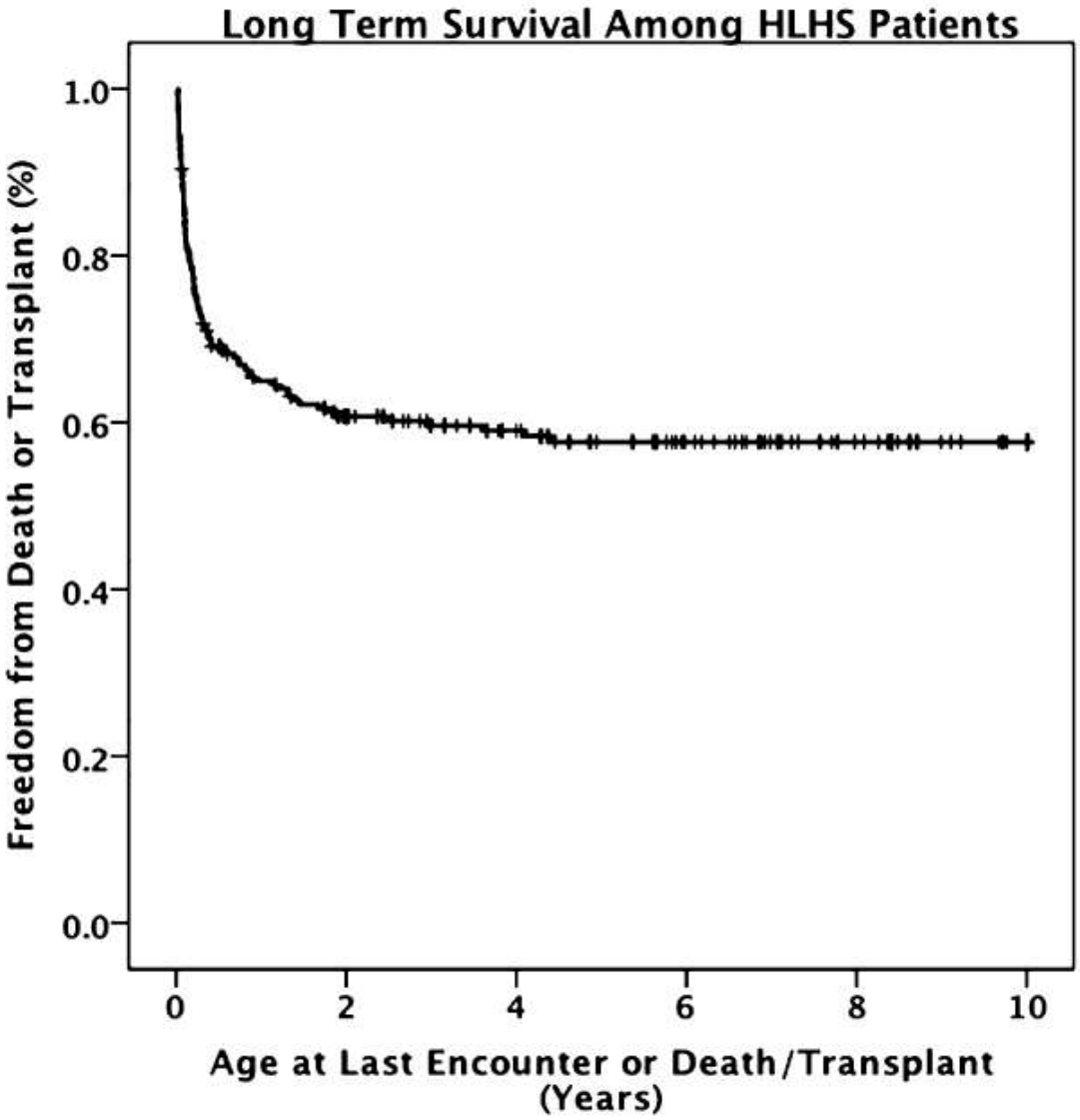


Figure 14-11 Damus-Kaye-Stansel operation for complete transposition of the great arteries (D-TGA) plus ventricular septal defect (VSD) plus subaortic stenosis.

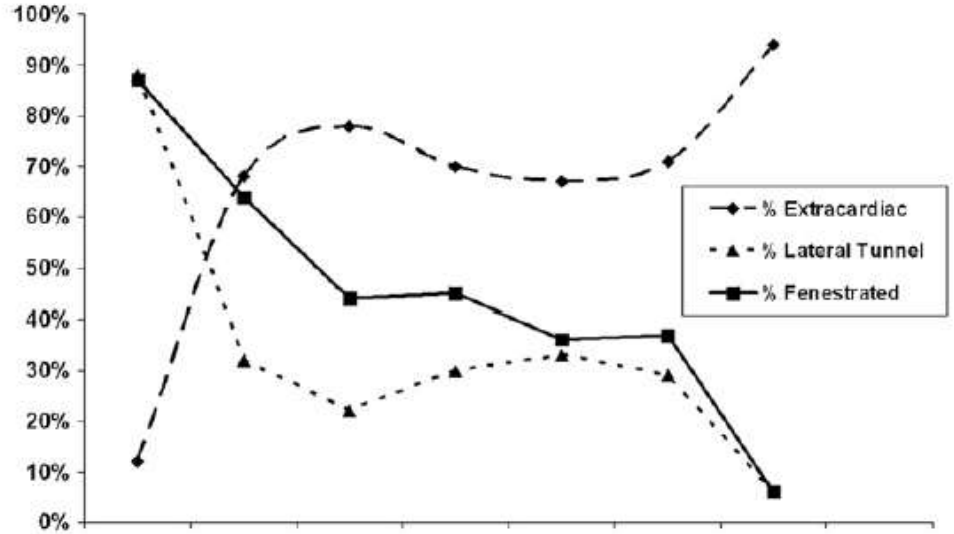
Variable	Low risk	Medium risk	High risk
PVR (Wood's units)	<2	2-4	>4
Mean PAP (mmHg)	<15	15-20	>20
LVEDP (mmHg)	<8	8-12	>12
EF (%)	>60	45-60	<45

PAP, pulmonary artery pressure; LVEDP, left ventricular end-diastolic pressure; EF, ejection fraction.


Long Term Survival Among HLHS Patients
Freedom from Death or Transplant (%)
1.0
0.8
0.6
0.4
0.2
0.0
0
2
4
6
8
10
Age at Last Encounter or Death/Transplant (Years)

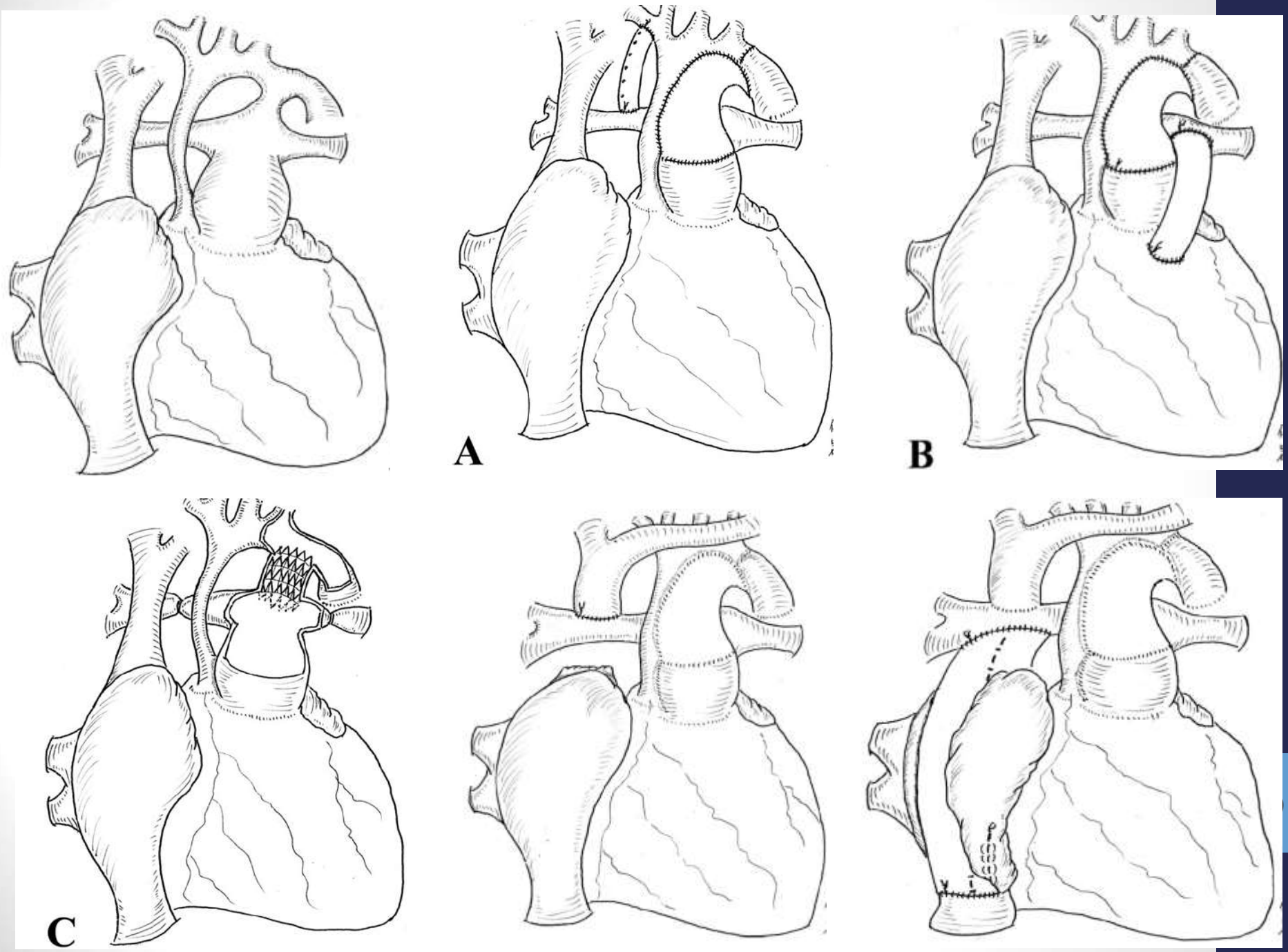
Fenestration during Fontan palliation: Now the exception instead of the rule

Jorge D. Salazar, MD,[a,d] Farhan Zafar, MD,[d] Kashif Siddiqui, MD,[d] Ryan D. Coleman, BA,[d] David L. S. Morales, MD,[a,d] Jeffrey S. Heinle, MD,[a,d] Joseph W. Rossano, MD,[b,e] Emad B. Mossad, MD,[c,f] and Charles D. Fraser, Jr, (J Thorac Cardiovasc Surg 2010;140:129-36)



	2002	2003	2004	2005	2006	2007	2008	Totals
Extracardiac Conduit	2 (50%)	21 (52%)	21 (33%)	28 (25%)	26 (11%)	29 (21%)	30 (0%)	157 (22%)
Lateral Tunnel	14 (93%)	10 (100%)	6 (83%)	12 (92%)	13 (85%)	12 (75%)	2 (100%)	69 (88%)
Totals	16 (87%)	31 (64%)	27 (44%)	40 (45%)	39 (36%)	41 (37%)	32 (6%)	226 (42%)

FIGURE 1. Relative use of extracardiac versus lateral tunnel technique and association with overall fenestration. Numbers of patients with percentages of fenestration are shown at *bottom*.


A
B
C

Hypoplastic Left Heart Syndrome

Surgical correction occurs in three stages:

Stage I: Norwood procedure

- Ascending aorta and arch reconstruction.
- PDA ligation.
- Construction of a reliable pulmonary blood flow source using a BT shunt or Sano shunt.
- Anesthetic management includes pre-bypass PGE_1 infusion, maintenance of nearly equal pulmonary and systemic blood flow for adequate systemic perfusion, precautions against air embolism, maintenance of anesthesia with intravenous drugs, and postbypass maintenance of a high hematocrit and probably a need for inotropic support.

Stage II: Glenn procedure

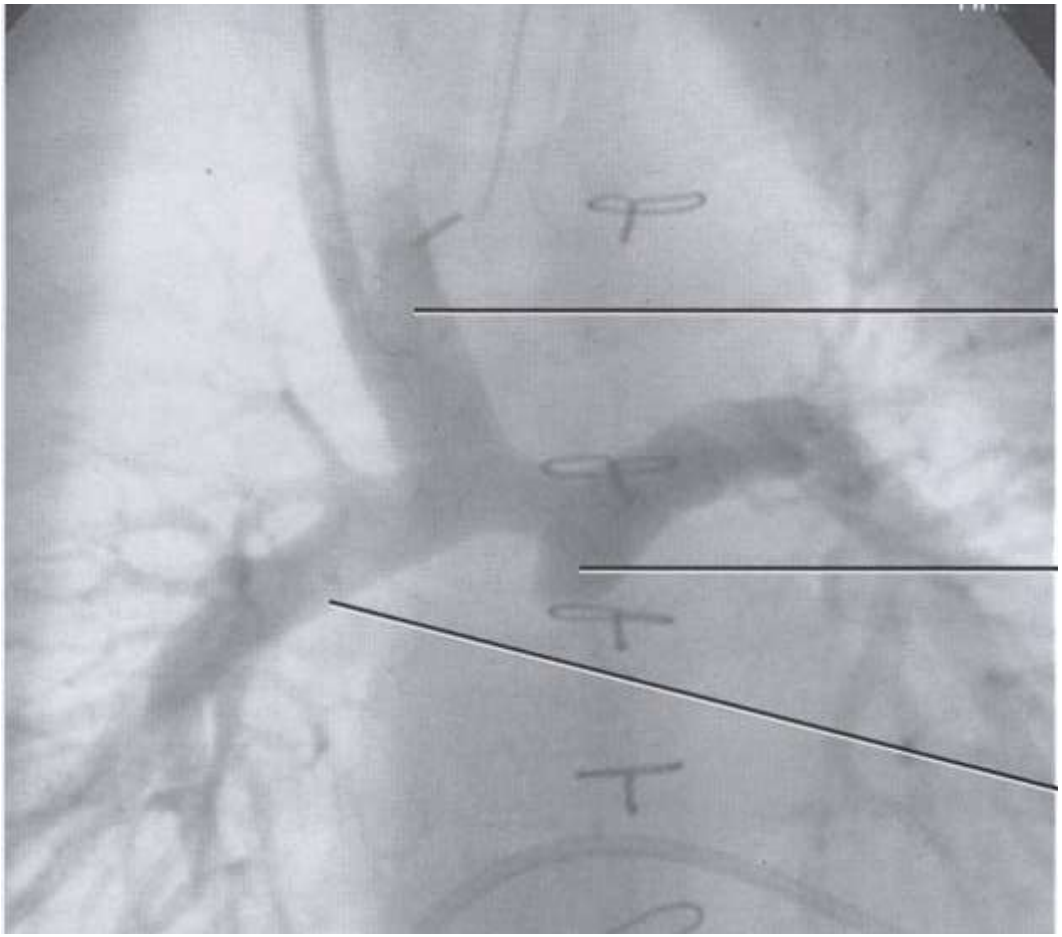
- Creation of a direct connection between the superior vena cava and pulmonary artery.
- Anesthetic management includes maintenance of a high hematocrit, elevation of the head of the bed to facilitate venous drainage, avoidance of central lines to reduce the risk for pulmonary artery thrombus and recognition that positive-pressure ventilation of the patient's lungs may decrease pulmonary blood flow and cardiac output.
- Mild hypoventilation may increase oxygen saturation.

Stage III: Fontan procedure

- Rerouting of blood flow from the inferior vena cava into the pulmonary circulation, usually accomplished using an extracardiac conduit.
- Preload to the heart is completely passive. Management of a patient status post Fontan procedure should focus on maintaining reasonable preload, i.e., passive flow from systemic veins to the pulmonary artery and eventually to the common atrium.
- Poor prognostic factors are high pulmonary vascular resistance, tricuspid regurgitation, and decreased ventricular function.

Tricuspid Atresia or Pulmonary Atresia with Intact Ventricular Septum

- Usually the right ventricle is diminutive or hypoplastic.
- Surgical approach involves an aortopulmonary shunt and subsequent Glenn and Fontan procedures.



Orijinal Fontan Operasyonu

1971

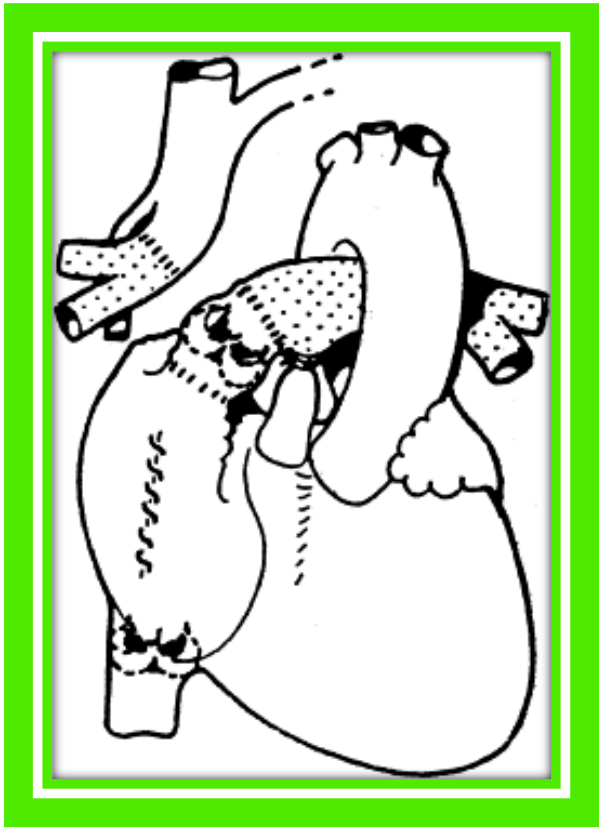
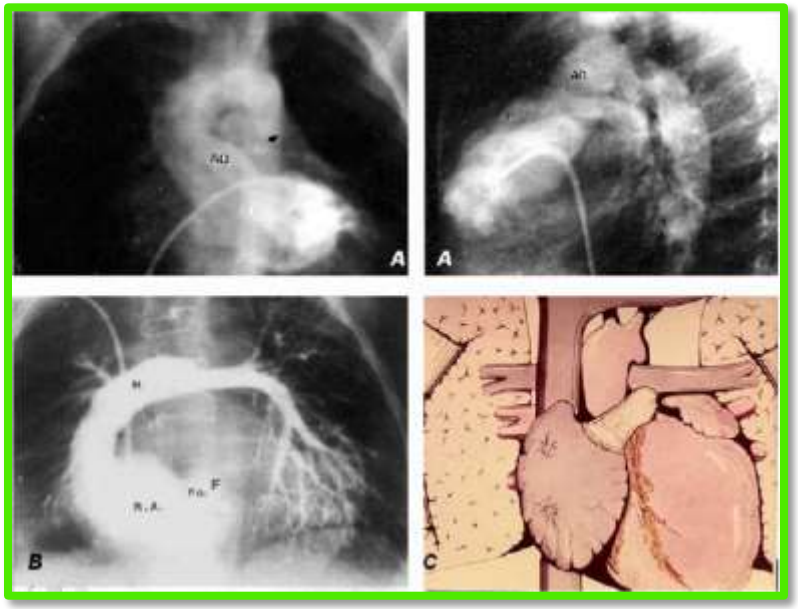


Fig. 1. The Fontan operation as originally described by F. Fontan for tricuspid atresia was comprised of five steps: (1) Glenn shunt to the right lung; (2) harvest of the main pulmonary artery from the right ventricular outflow tract; (3) anastomosis of main pulmonary artery to the right atrium with an interposed homograft valve; (4) placement of a homograft valve in the inferior vena cava-right atrium junction; and (5) closure of atrial septal defect. (Reprinted with permission from Fontan F, Baudet E, Thorax 1971;26:240.)

Kreutzer Modifikasyonu APC

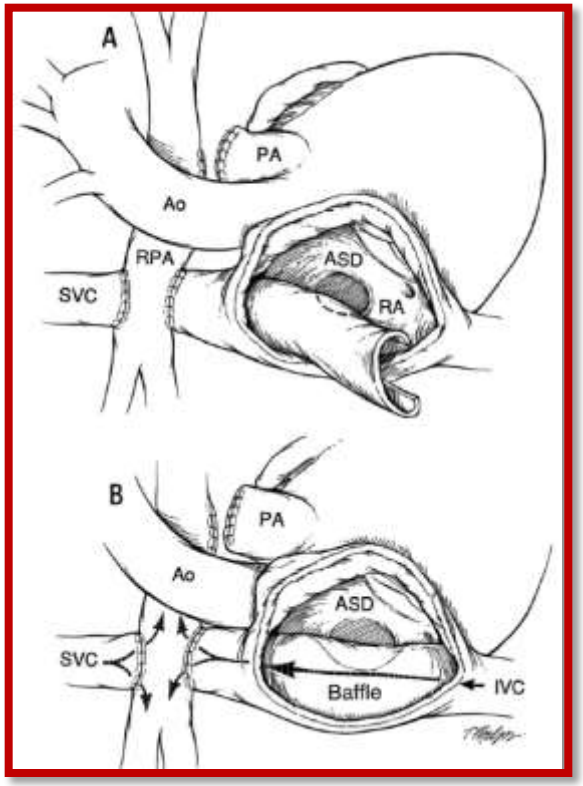
1973-1982



Homogreft ve kapak kullanmadan direkt anastomozu tanımlıyor

De Leval - TCPC

1988



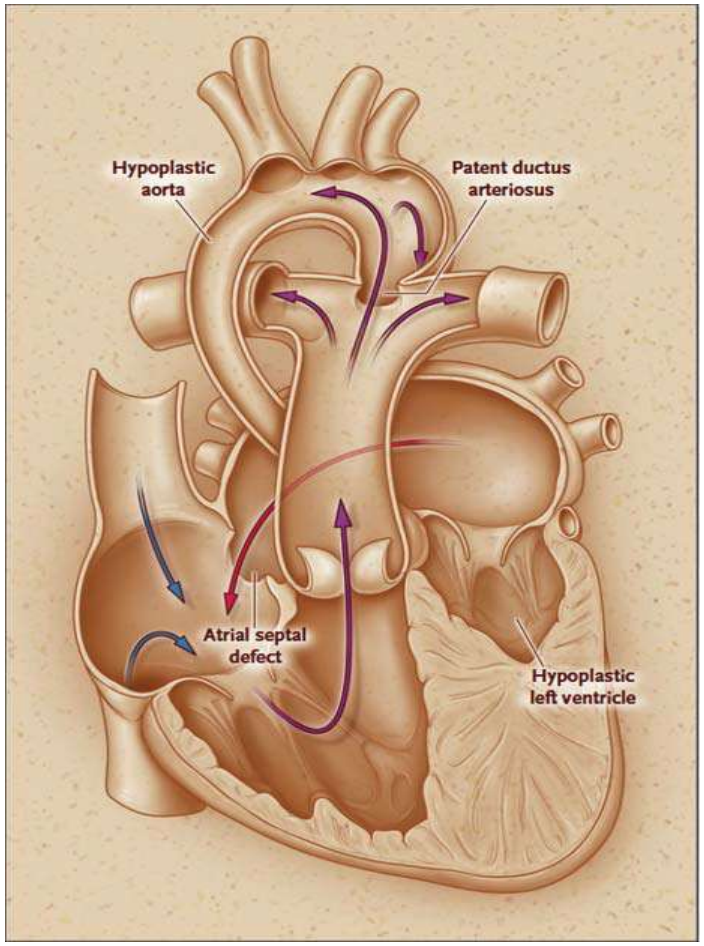
Lateral Tünel

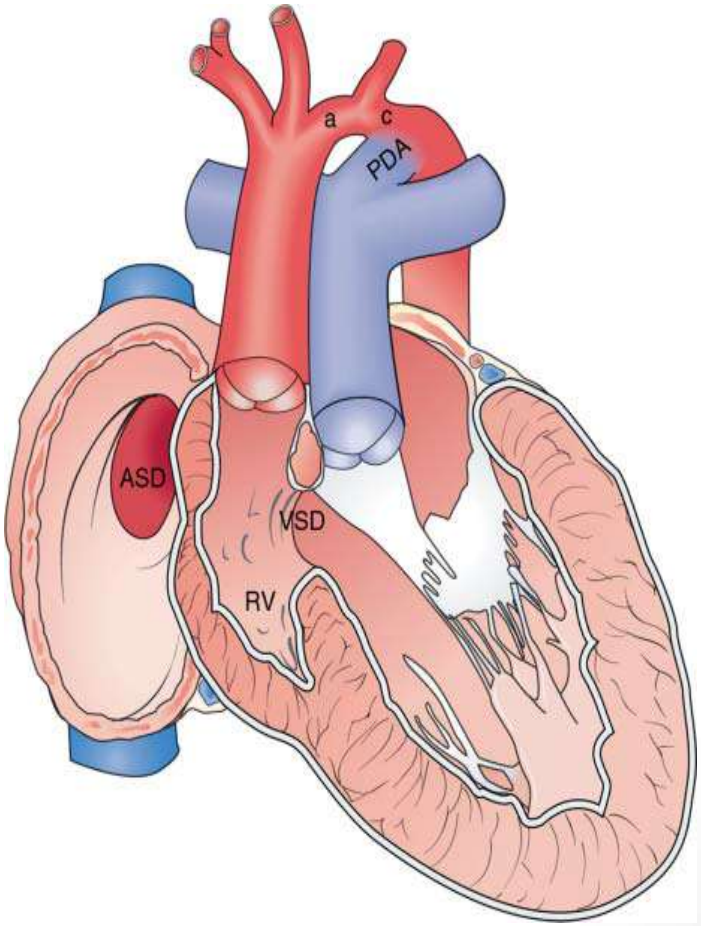
- *Dacron veya PTFE ile atriumun bölünmesi*
- *Düz bir yol oluşturulması*
  - *Türbulans ve enerji kaybı olmadan laminar akım sağlanması*

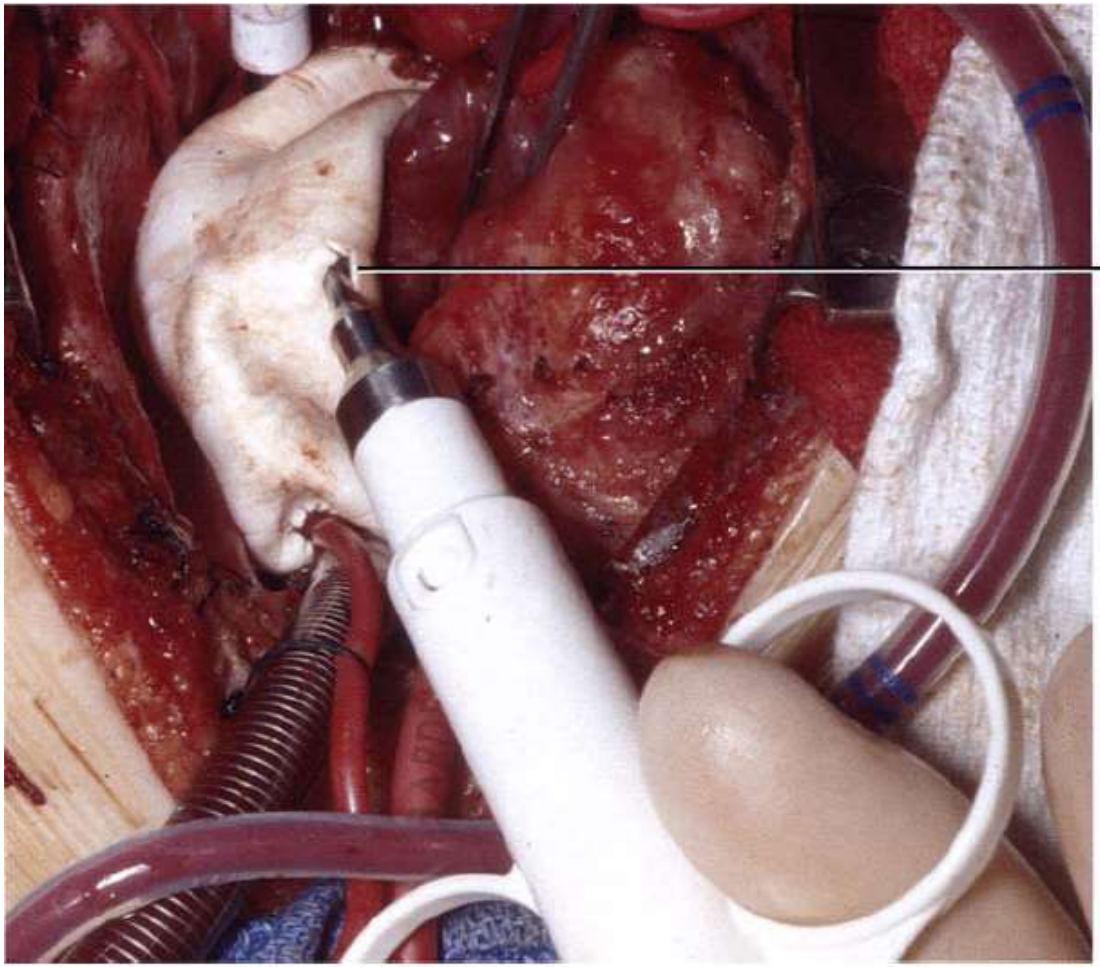
Tek ventrikül Anatomisi

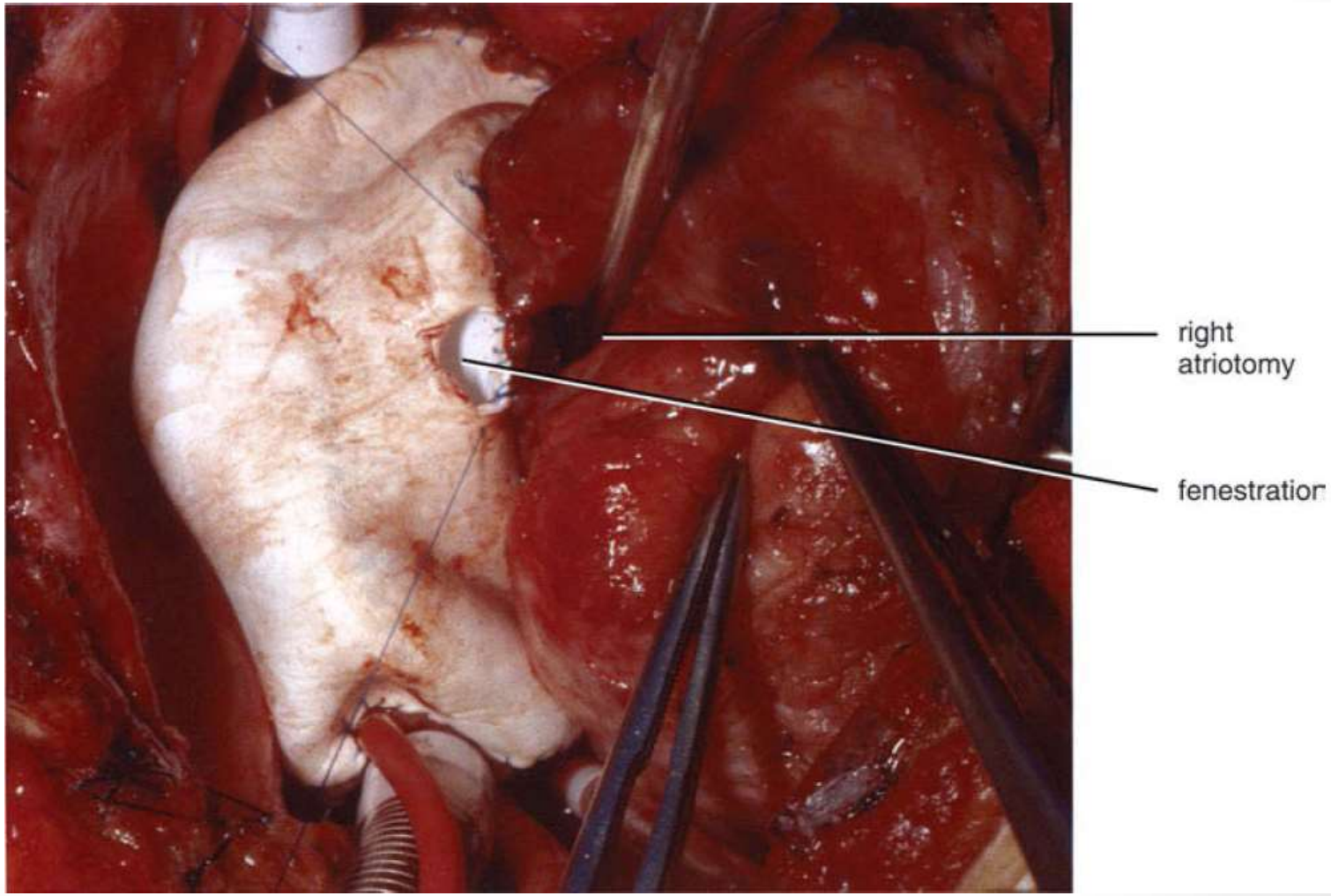
- Pulmoner kan akımı norml ancak sistemik kan akımı obstrüksiyonu
- (2) Normal sistemik kan akımı ancak obstrükte akciğer kan akımı;
- (3) Bilateral engelsiz kan akımına ilave olarak,Ya sistemik veya pulmoner venöz dönüş de de obstrüksiyon olabilir

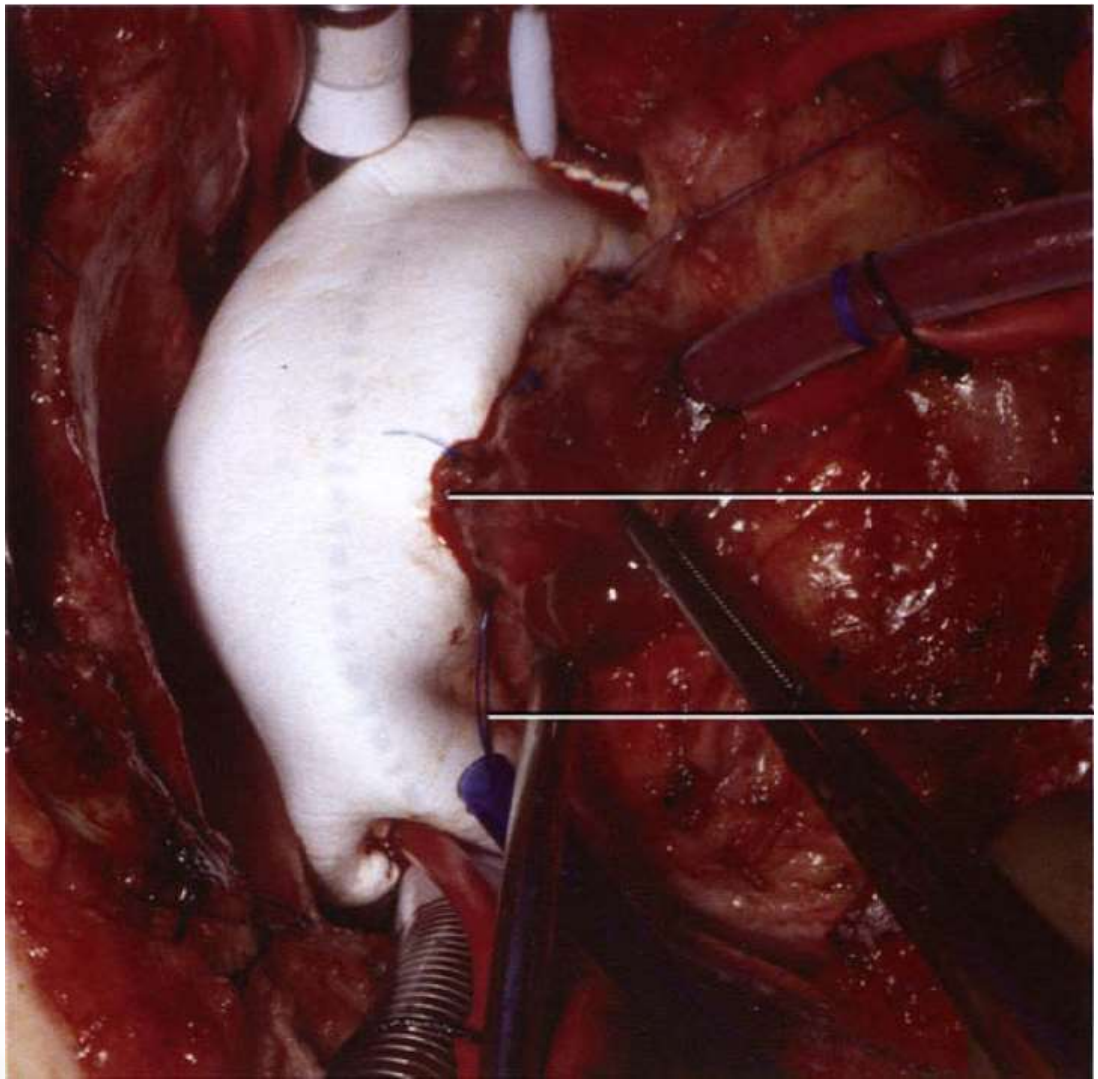
En sık karşılaşılan Morfolojisi





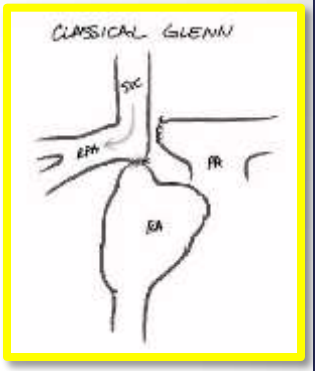

fenestration


right
atriotomy
fenestration




fenestration

Glenn ameliyatı



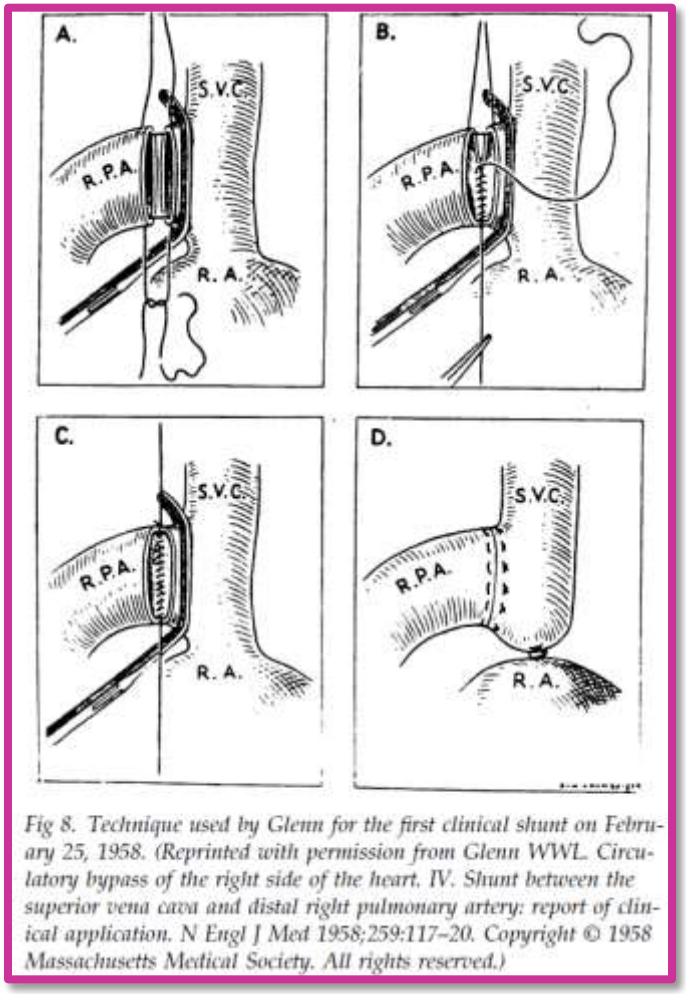


Fig 8. Technique used by Glenn for the first clinical shunt on February 25, 1958. (Reprinted with permission from Glenn WWL. Circulatory bypass of the right side of the heart. IV. Shunt between the superior vena cava and distal right pulmonary artery: report of clinical application. N Engl J Med 1958;259:117–20. Copyright © 1958 Massachusetts Medical Society. All rights reserved.)

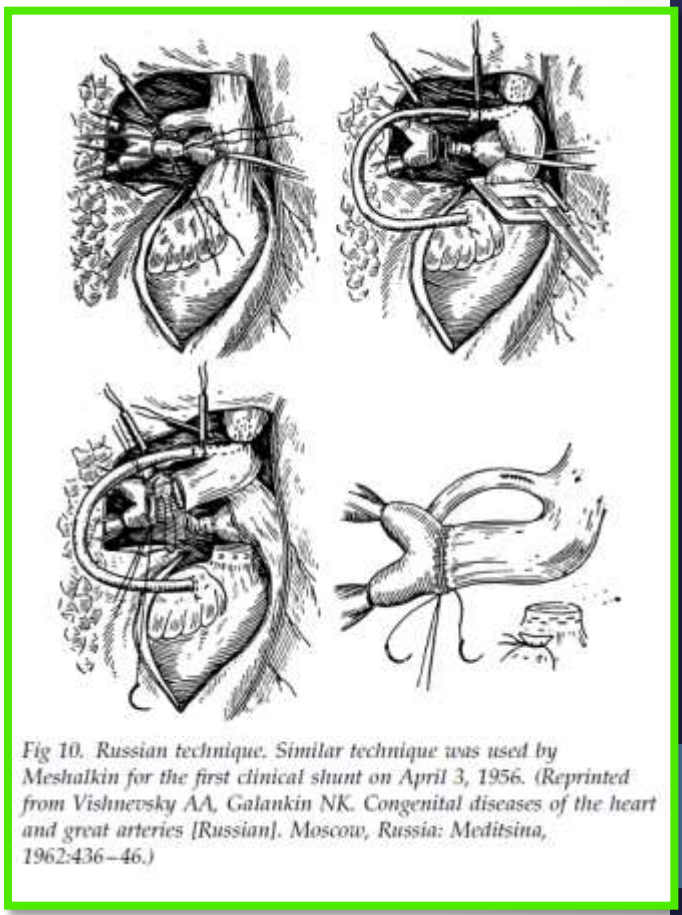
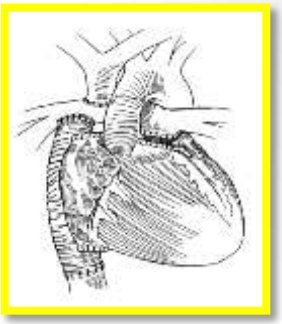


Fig 10. Russian technique. Similar technique was used by Meshalkin for the first clinical shunt on April 3, 1956. (Reprinted from Vishnevsky AA, Galankin NK. Congenital diseases of the heart and great arteries [Russian]. Moscow, Russia: Meditsina, 1962:436–46.)

Ekstrakardiak Fontan

1988-1990



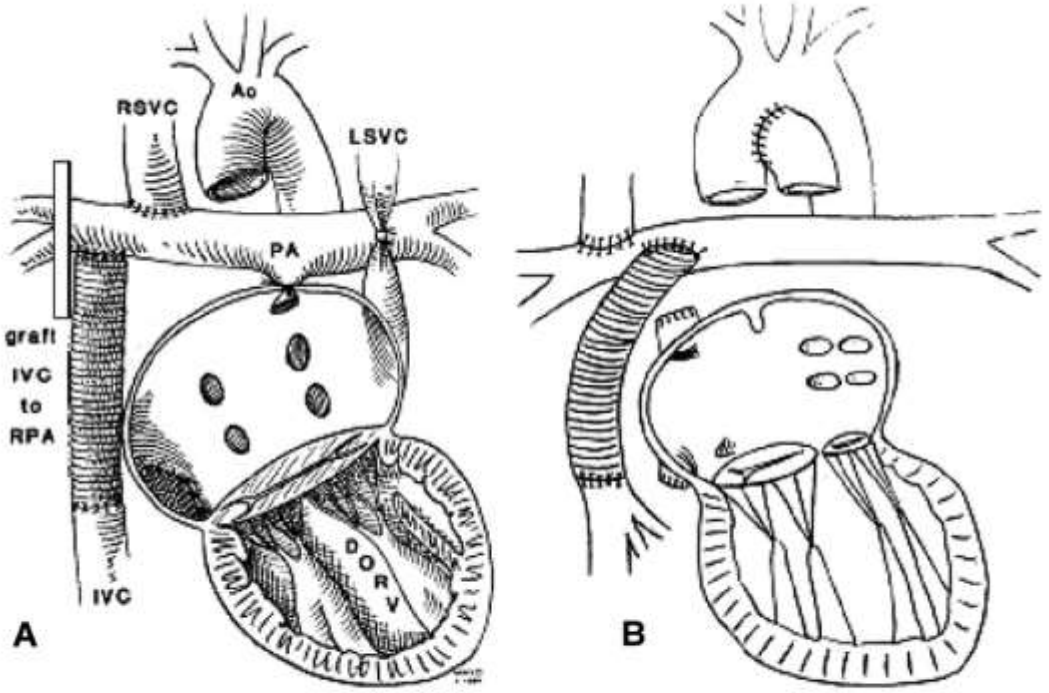


Figure 5 A, EC, performed by Puga.[14] (Reprinted with permission.) B, EC, performed by Marceletti.[15] (Reprinted with permission.)

KEY POINTS: SUPERIOR CAVOPULMONARY ANASTOMOSIS

- SCPA is performed at 3–6 months of age and is normally the second stage of palliation in the Fontan pathway.
- SCPA results in significant unloading of the systemic ventricle and stabilization of the circulation, particularly in HLHS.
- Surgery is done with CPB but without aortic cross-clamping unless additional intracardiac surgery is required.
- The creation of a "cavopulmonary–cerebral" circulation with SCPA is an important concept; mild hypercarbia will promote cerebral and cavopulmonary blood flow and improve cerebral and systemic oxygenation and oxygen delivery.

Fenestrasyon rutin veya riskli vakalarda ?

- *Tek AC Fontan*
- *Yüksek pulmoner arter basıncı*
- *Yüksek pulmoner vasküler rezistans*
- *Anormal pulmoner arter anatomisi*
- *Sağ ventrikül dominansı*
- *Bozuk ventrikül fonksiyonu*
- *Belirgin AV kapak yetmezliği*
- *Erken cerrahi*

Basınça göre değişen fenestrasyon

- *2.5 mm altında fenestrasyonda tromboemboli riski çok düşük*
- *İhtiyaç halinde (sistemik venöz ile pulmoner venöz atrium arasında P artması ile) ~8-10 mmHg*
- *Fenestrasyon çapı artarak hemodinami rahatlıyabilirmi ?*

Fenestrasyonun seyri

- *2.5-4 mm=== %50-90* *1 yılda kapanıyor*
- *4 mm > === %40-70* *1 yılda kapanıyor*

Reported survival rates following Fontan operation in recent series range from 86% to 94% at 10 years (100,113-116), and 82% to 87% at 15 to 20 years (100,101,117). However, if PLE develops, the five-year survival is approximately 50% (95). The most common causes of late death are arrhythmias, heart failure and thromboembolic complications (118).

CHD is a risk factor for adverse outcome after heart transplantation (119), with reported survival rates of 72% to 86% at one year, 68% to 77% at five years and 62% at 10 years (120-122).

Part II. History and management of unoperated patients

The prognosis of all patients with unoperated univentricular hearts is poor, with a median survival of 14 years (death rate of 4.8% per year). The majority are symptomatic with cyanosis and exercise intolerance (131).

ikinci asama

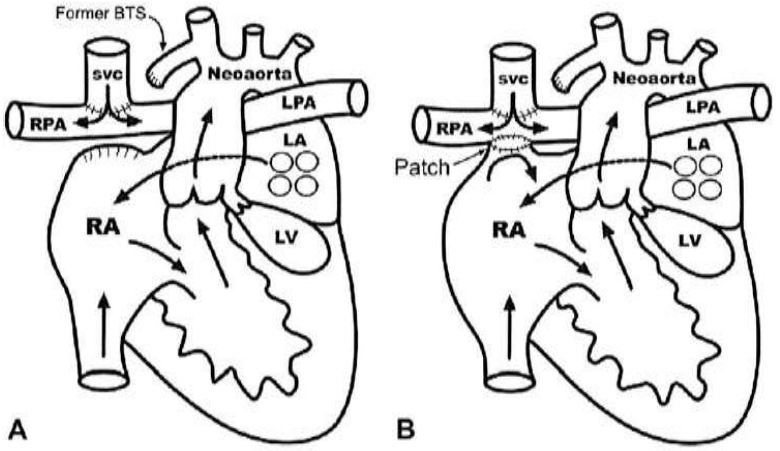
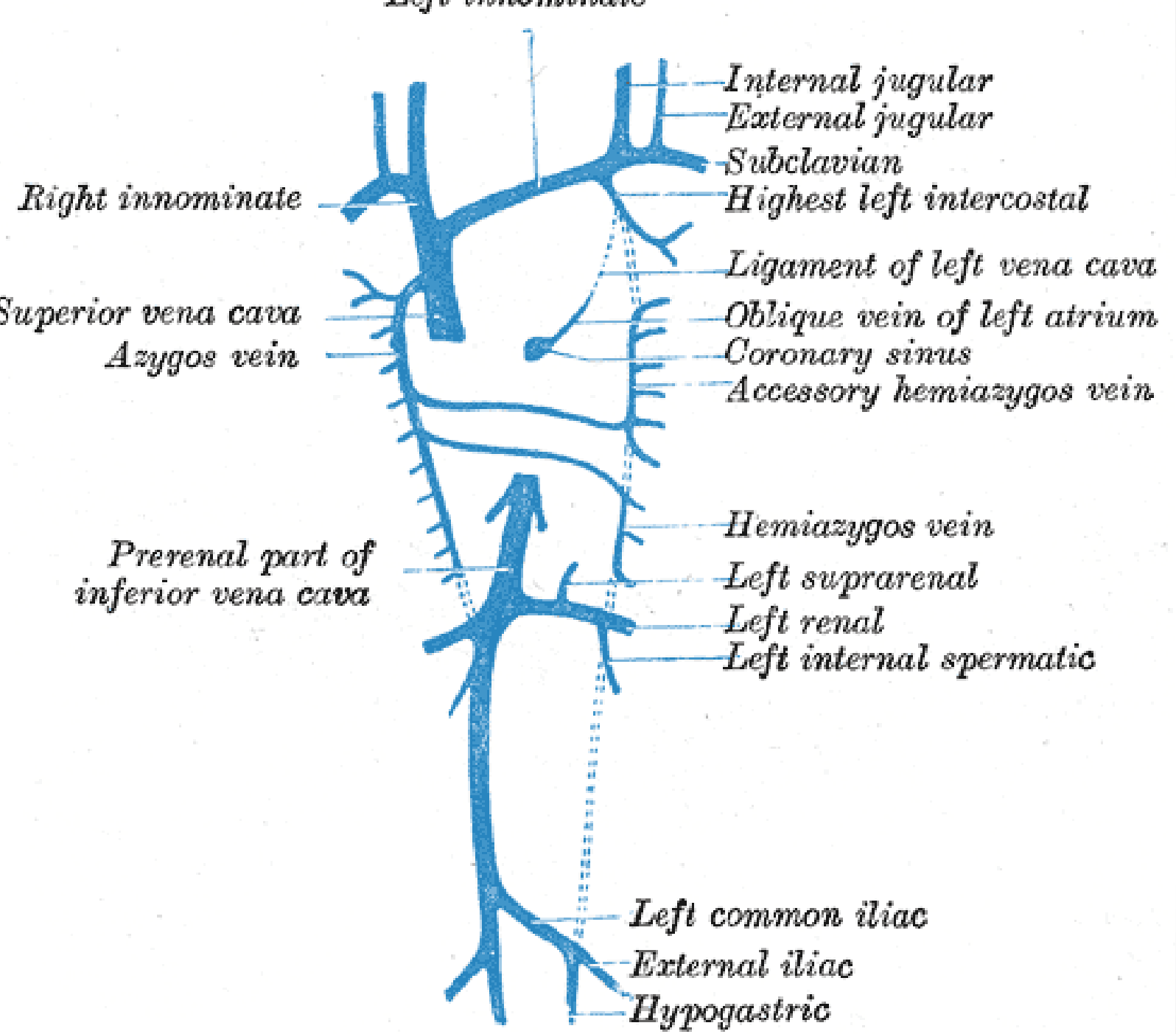


Fig. 3. Technical variants of stage II partial cavapulmonary anastomosis, immediate postoperative period. Note the ventricular dilation and hypertrophy caused by volume overload during the preceding stage. (*A*) The bidirectional Glenn. (*B*) The hemi-Fontan. This procedure is physiologically identical to the Glenn, but with an additional anastomosis from the proximal SVC to the RPA. This anastomosis is then occluded with a patch, which is removed during subsequent Fontan completion. Direction of blood flow indicated by the *arrows*. BTS, Blalock-Taussig shunt; LA, left atrium; LPA, left pulmonary artery; LV, left ventricle; RA, right atrium; RPA, right pulmonary artery; RV, right ventricle; SVC, superior vena cava. (*Reprinted from* Walker SG, Stuth EA. Single-ventricle physiology: perioperative implications. Semin Pediatr Surg 2004;13(3):195; with permission from Elsevier.)


Left innominate
Internal jugular
External jugular
Subclavian
Right innominate
Highest left intercostal
Ligament of left vena cava
Superior vena cava
Oblique vein of left atrium
Azygos vein
Coronary sinus
Accessory hemiazygos vein
Hemiazygos vein
Prerenal part of inferior vena cava
Left suprarenal
Left renal
Left internal spermatic
Left common iliac
External iliac
Hypogastric

İlk Aşama: Yenidoğan palyasyon II

Sistemik çıkım obstrüksiyonu:

- Aorta koarktasyonu
  - Torakotomi
  - Sternotomi HSA
  - Diğer kanülasyon teknikleri SSP için
- Mortalite %...

İlk Aşama: Yenidoğan palyasyon III

Sistemik Çıkım obstrüksiyonu

- Hipoplastik sol kalp veya ciddi ve düzeltilemez sistemik çıkım obst olan unbalance ventrikül olguları: Norwood, Sano or Damus–Kaye–Stensel type intervention
- Mortalitenin yüksek olacağı yada düzeltme yapılamayacak olgular,
  - Hybrid yaklaşım or orthotopic heart transplant

İlk Aşama: Yenidoğan palyasyon IV

- yüksek pulmoner kan akımı ile birlikte Pulmoner arteriyel hipertansiyon olgularında ise Pulmoner artere bant konur.

Birinci asama: YeniDogan: Hipoplastik sol kalp Sistemik Çıkım obstrüksiyonu: HİBRİD

- HLHS ile 323 yenidoğan çok merkezli çalışma
- 30 günlük mortalite her iki yaklaşım için % 33 oranı
  - (Nakil veya kademeli palyasyon)

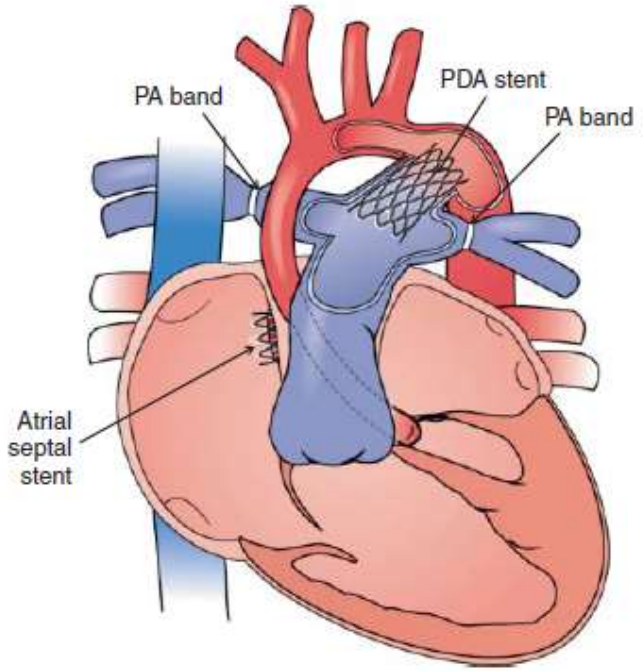
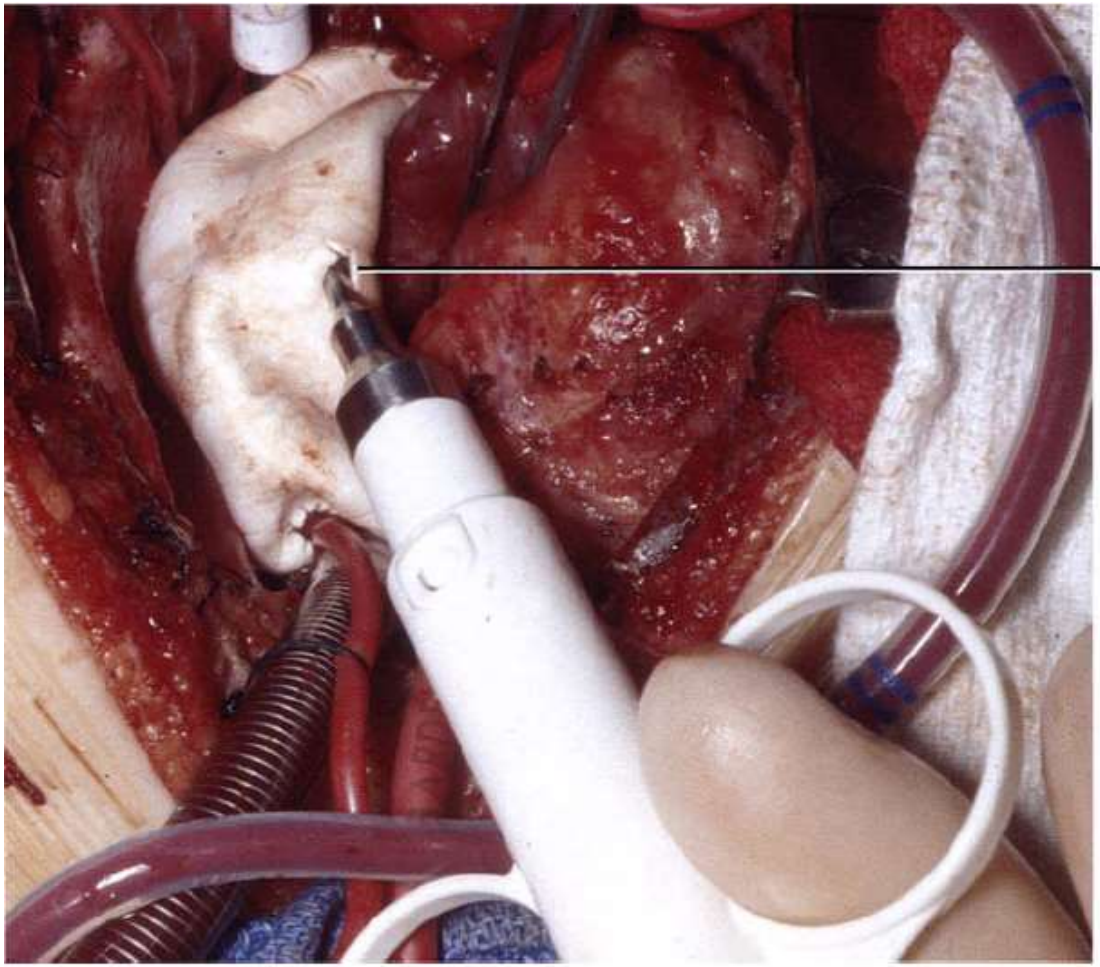
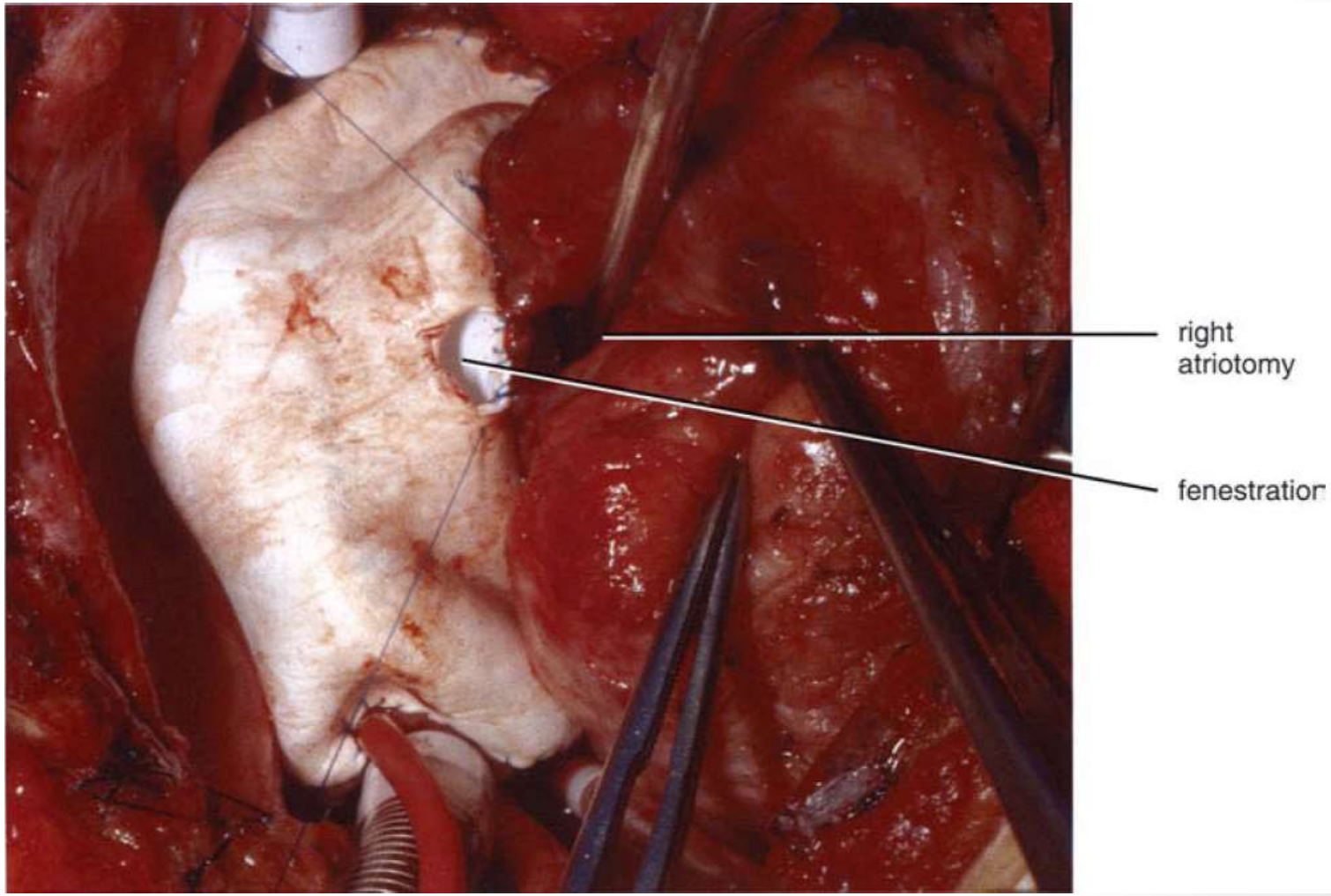
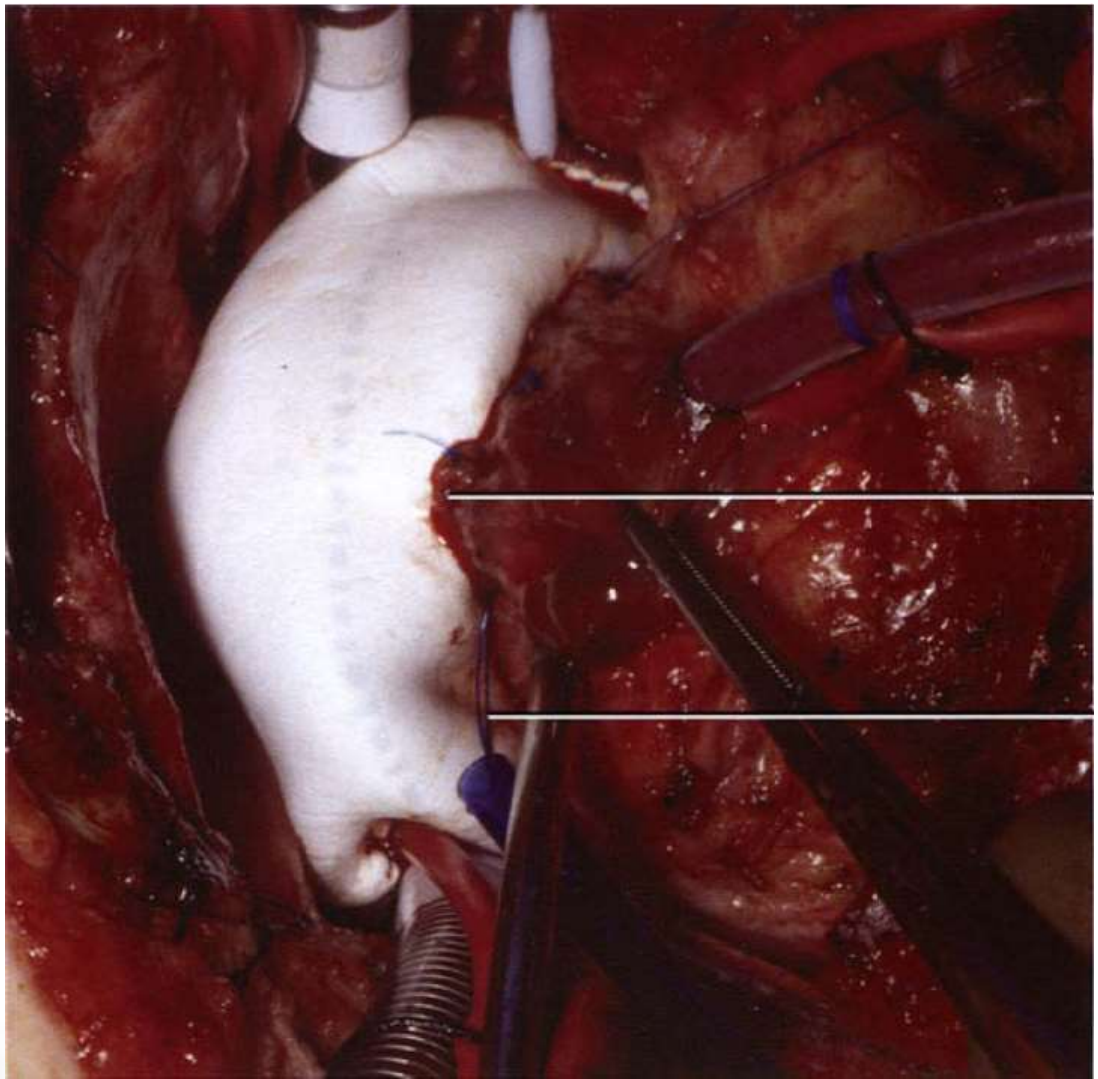


Figure 25.5 The hybrid stage I palliation for hypoplastic left heart syndrome. A median sternotomy is performed, and without bypass, bilateral pulmonary bands are placed, and a stent is placed in the patent ductus arteriosus via a sheath in the main pulmonary artery positioned by the interventional cardiologist. A stent is shown in the atrial septum. PA, pulmonary artery; PDA, patent ductus arteriosus. (Source: Andropoulos & Gottlieb [20]. Reproduced with permission of Elsevier.)


fenestration


right
atriotomy
fenestration



Tek ventrikül ve kalp nakli

- Bu strateji HLHS ile yenidoğanlar için önerildiğinde oldukça taraftar kazandı
- yıllık olarak 500 pediatrik kalp nakli dörtte biri 1Yaşından küçük
- bebeklerde kalp nakli yapılan hastalarda 10 yıllık sağkalım oranları % 76 olarak bildirilmiştir.

özet

- Tek ventrükül lezyonu tüm yd konj anomalilerin %5ini oluşturur
- Bu hastalar stage palyatif rekonstriksiyona giderler.
- Bir çok nedenle tekrar tekrar anestezi alırlar
- Optimal sonuçlar dikkatle anesteziyologlar, cerrahlar, kardiyologlar ve Yoğun bakım arasındaki etkileşimin düzenlenmesini gerektirir

Fontan: geç Sonuçlaroutcame

- Significant morbidity and late mortality can occur following Fontan completion, including deteriorating ventricular function, arrhythmias, thrombotic events, and protein-losing enteropathy (PLE).
- In a cohort of 330 patients evaluated a median of 8 years following Fontan operation, the rates of freedom from death or transplant at 5 and 10 years were 95% and 93%, respectively.
- school performance was above average (30%) or average (40%
- Sequelae of the Fontan operation include arrhythmias %25-50, thrombotic events (%10), and PLE (%3.8).
- PLE protein kaybettiren entoropati: tıbbi veya cerrahi müdahaleler son derece dirençlidir ve 5 yıl mortalite % 50.
- Plastik bronşit Fontan operasyonu [118] sonra nadir fakat ciddi ve genellikle hayatı tehdit eden sekeller olduğun

arter cardiac surgery

MD; Michele Merat, MD; Gustavo La Rotta, MD; Pretha Joshi, MD; Vinay Joshi, MD;
Steve Jarvis, RT; Christopher A. Caldarone, MD; Glen S. Van Arsdell, MD;
ngton, MD; Brian P. Kavanagh, MD

othesized that spontaneous inspiratory effort
leural space during airway pressure release
sult in increased lung perfusion after surgery
ot or following a cavopulmonary shunt as a
sient decreases in intrapleural pressure.
ive crossover cohort study.
y care cardiac pediatric intensive care unit.
n after tetralogy of Fallot repair, cavopulmo-
n operation.
ng perfusion and cardiac output were mea-
y pressure release ventilation and pressure
ith pressure support, both with and without
tion. Oxygen consumption was measured
) and lung perfusion/cardiac output calcu-
). Constant levels of CO_2 and mean airway
ted in all study phases.
nd Main Results: Twenty patients were en-
nine after repair of tetralogy of Fallot and 11
ary shunt. In the absence of spontaneous ventilation, there were no differences in lung perfusion or any of the measured gas exchange or hemodynamic parameters. In the presence of spontaneous ventilation for all patients, mean pulmonary blood flow increased from 2.4 to 2.9 $L \cdot min^{-1}M^{-2}$ ($p = .02$). Oxygen delivery increased from 594 to 774 $mL/min/m^2$ ($p = .05$) in the patients with tetralogy of Fallot patients and from 473 to 518 $L \cdot min^{-1}M^{-2}$ ($p = .07$) in the cavopulmonary shunt group.

Conclusion: Ventilation with airway pressure release ventilation (at comparable mean airway pressure) improves lung perfusion compared with pressure control ventilation in children after tetralogy of Fallot repair and cavopulmonary shunt operations. Although this study focused on tetralogy of Fallot and cavopulmonary shunt operations, the improved cardiopulmonary interactions may be beneficial in other situations in which hemodynamics are impaired by positive pressure ventilation. (Crit Care Med 2011; 39:2599–2604)

KEY WORDS: APRV; ventilation; cardiac surgery; tetralogy of Fallot; Fontan operation

ntraendotracheal pressure for a patient during airway pressure release ventilation
using periodic decreases in intraendotracheal pressure can be seen.

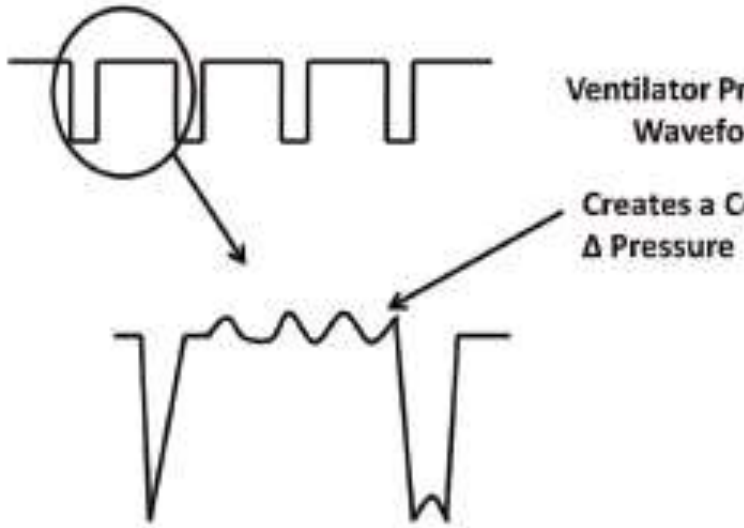
Ventilator Mandate

vay Pressure

Pleural Pressure

essure control ventilation with
ressure can be seen.

Airway Pressure Release Ventilat



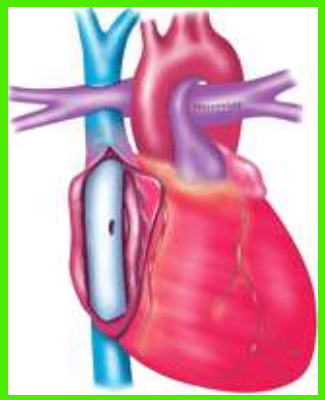
= ↑ Venous Return + ↓ RV Afterload

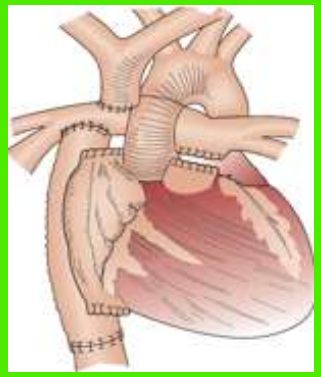
Figure 4. This schematic shows how syn
nized ventilation can negate any beneficial
of spontaneous respiration. We can see ho
way pressure release ventilation benefits p
nary blood flow by creating a constant osci
of pressure in the chest. *RV*, right ventricl

Fontan Ekstrakardiak ? Lateral ?

- Daha fazla laminar akım
- Atriyum insizyon ve sütür az
- Atrium basınç az
- Atriumda yabancı materyel yok
- Baffle leak ihtimali yok
- Koroner sinüs düşük P venöz atrium tarafında kalır
- Kardioplejik arrest gerekmez
- Fenestrasyon ilavesi veya revizyonu CPB'suz.
- Venöz dönüş anomalisinde kolay
- Tranplantasyonda pulmoner arter rekons daha kolay

- Antikoagülasyon
- PAV Malformasyon riski yüksek
- Büyük greft yerleştirme zorluğu





TCPC sonrası uzun dönemde komplikasyonların Fizyopatolojisi

- Sistemik venöz basıncın artması ve pulsatil akımın kaybolması
- Ventrikül fonksiyonlarının bozulması
- Pulmoner venöz obstrüksiyon
- AV kapak yetmezliği
  - Unbalansa CAVSD sık
- Aritmiler
  - Atrium büyümesi, insizyonu ve dikişler ile atriopulmoner anastomozlar >lateral tünel >EKF
- İlerleyici Siyanoz
  - SVP yüksekliği ile çeşitli kollateral ve AVM