DOĞUŞTAN KALP HASTALIKLARI

(SOLDAN SAĞA GEÇİŞLİ)

Dr. Ercan Tutar

Doğuştan Kalp Hastalıkları

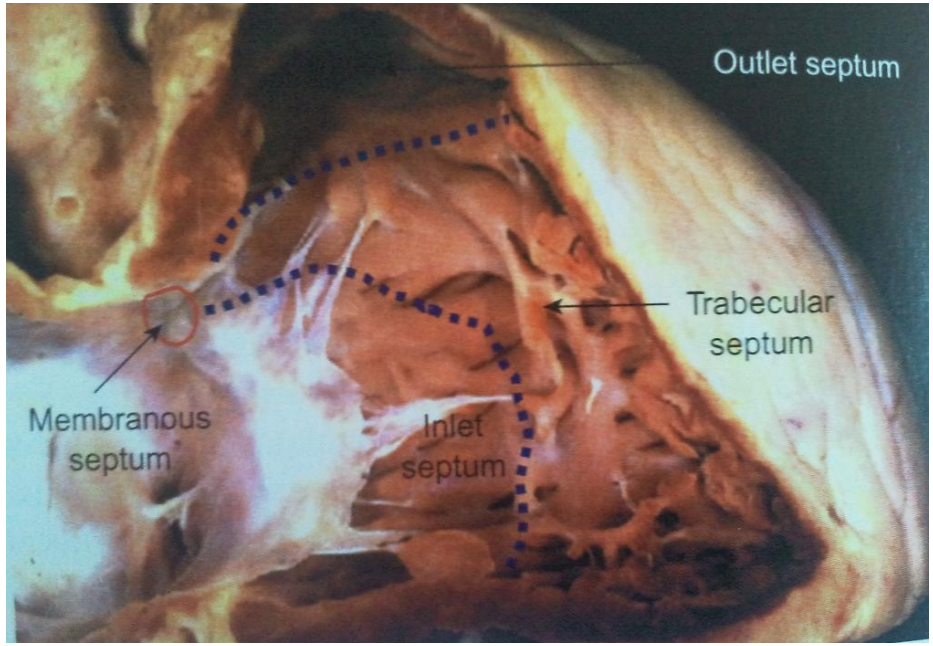
- **DKH sıklığı → 0.8 /100 canlı doğum**

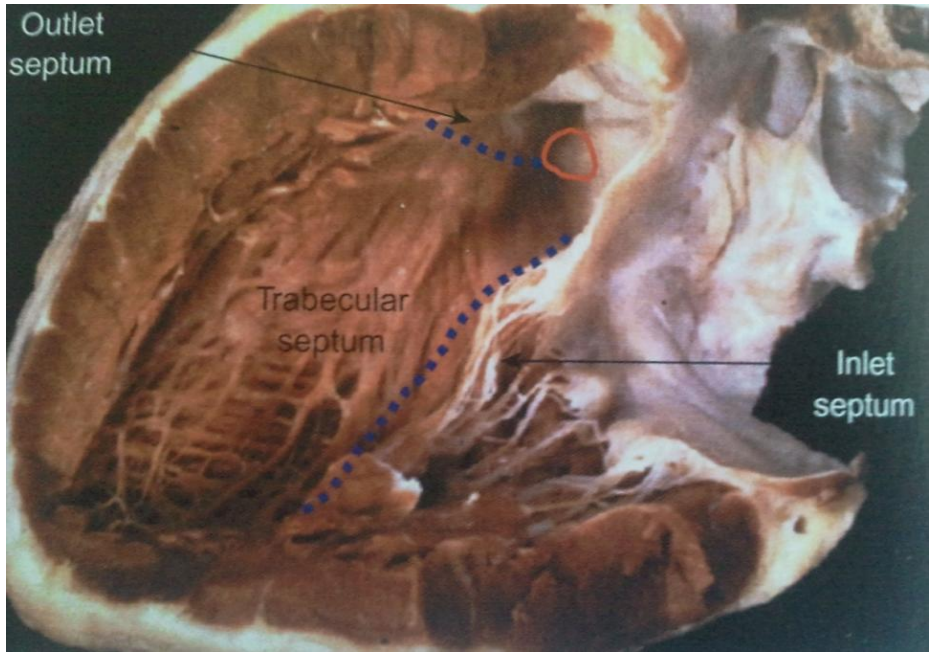
– **Soldan sağa geçişli DKH en sık**

- **Ventriküler septal defekt (VSD)**
- **Atriyal septal defekt**
- **Patent (persistan) duktus arteriyosus (PDA)**
- **Atriyoventriküler septal defekt (AVSD)**

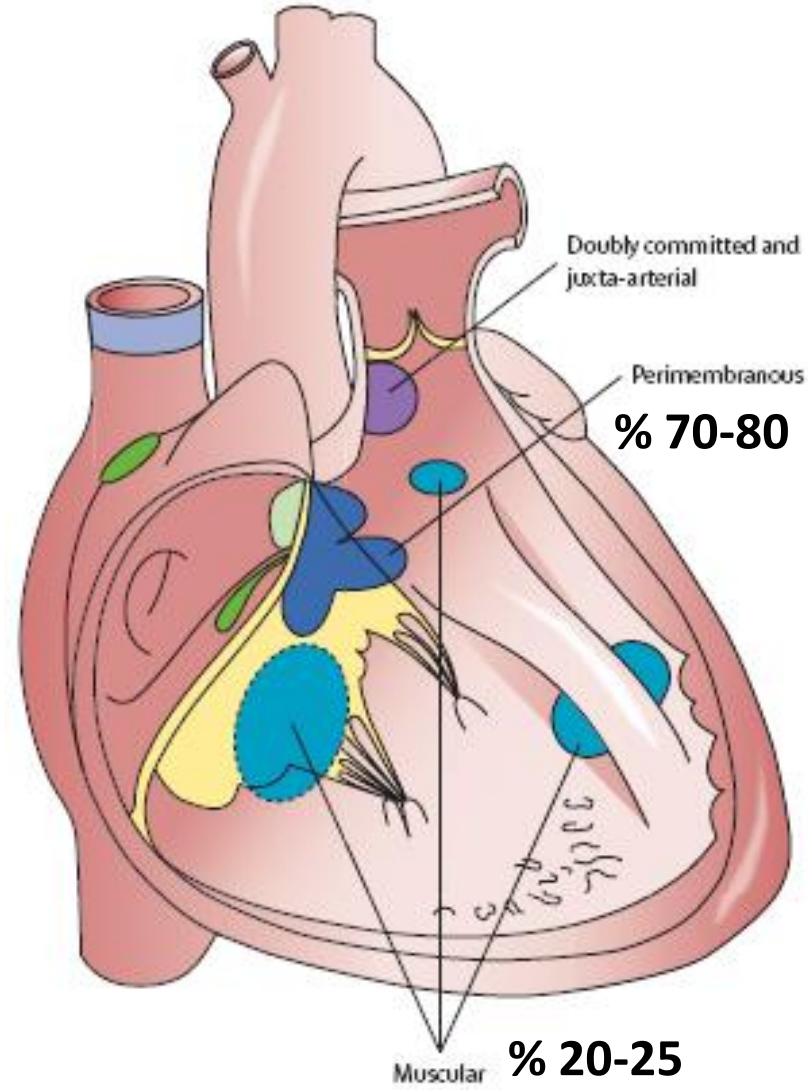
Ventriküler Septal Defekt
VSD

En sık görülen DKH → **% 20**





Echocardiography in Pediatric and Congenital Heart Disease, 2009



Lancet 2011; 377: 1103–12

VSD’de geçişin miktarını belirleyen etkenler

1. **Defektin büyüklüğü**
2. **Pulmoner damar direncinin derecesi**

Küçük VSD

Defekt sınırlayıcı

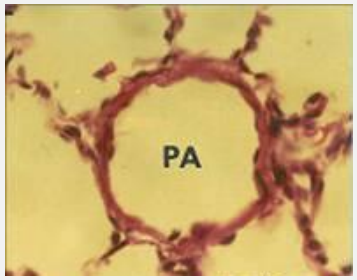
Geçiş PDD’ye bağlı değil

Büyük VSD

Defekt hiç sınırlayıcı değil

Geçiş başlıca PDD’ye bağlı

PDD düşük → **Geçiş fazla**



Orta-geniş VSD

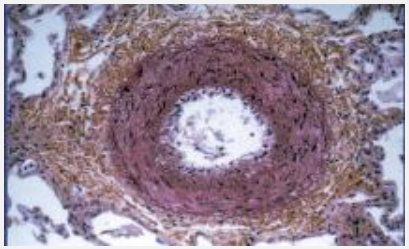
Artmış pulmoner kan akımı

(Damar duvarı gerilimi)

Basınç = AKIM x direnç

Endotel işlev bozukluğu ve damar duvarında yeniden yapılanma

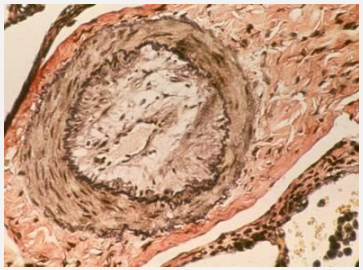
Vazokonstriksiyon, düz kas hücresi çoğalması, intimada değişiklikler, ekstrasellüler matriks artışı, fibrozis, intravasküler tromboz



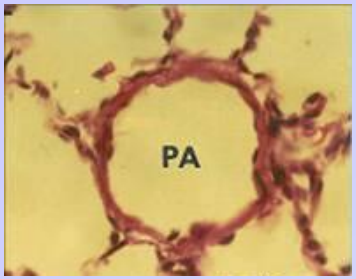
Pulmoner damar direnci artışı

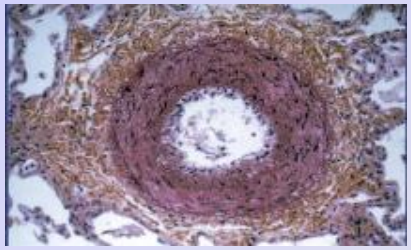
Basınç = akım x DİRENÇ

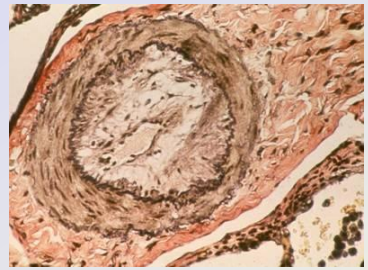
Şantın ters dönmesi



Eisenmenger sendromu







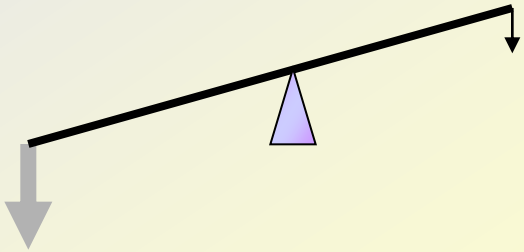
Prepro ET-1 → Big ET-1 (ECE) → ET-1 → ET_A, ET_B → Vazokonstriksiyon proliferasyon

L-arginin → L-sitrülin → Nitrik oksit → cGMP → (-) PDE5 → Vazodilatasyon Antiproliferasyon

Arakidonik asit → Prostasiklin → cAMP → Vazodilatasyon Antiproliferasyon

vazokonstriksiyon

proliferasyon

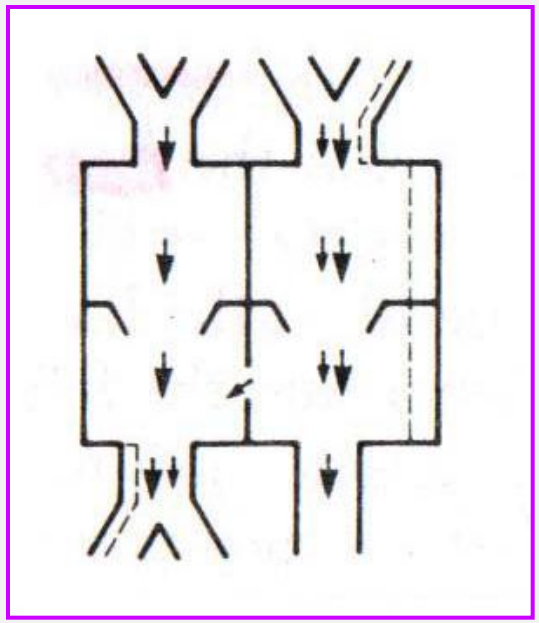


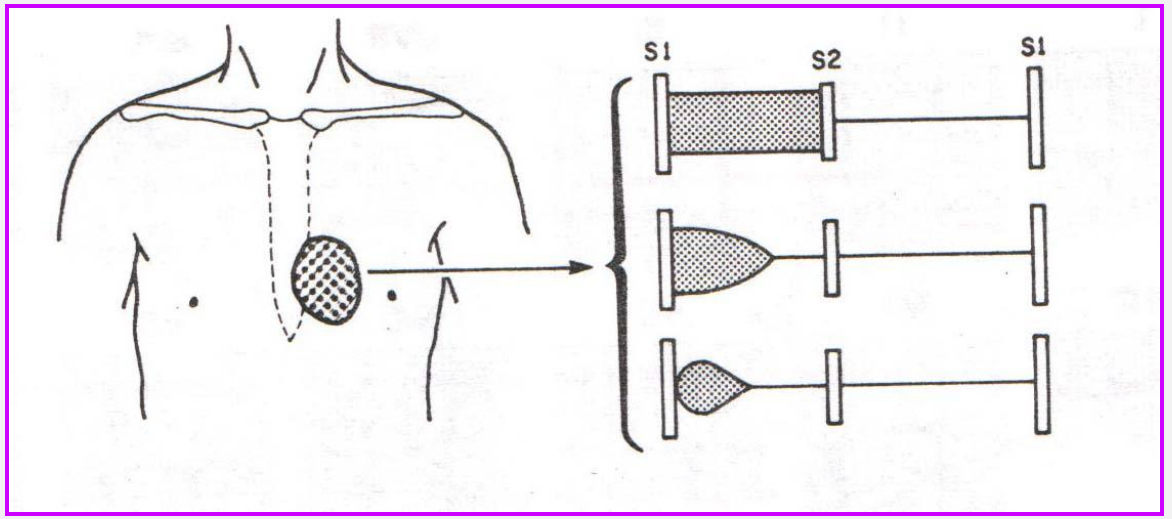
vazodilatasyon

antiproliferasyon

(J Am Coll Cardiol 2004;43:13S–24S)

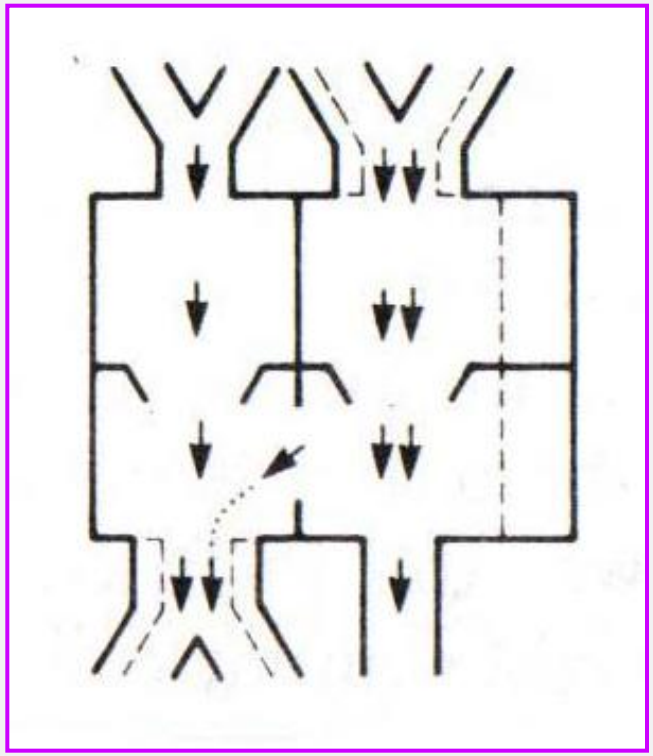
Küçük VSD

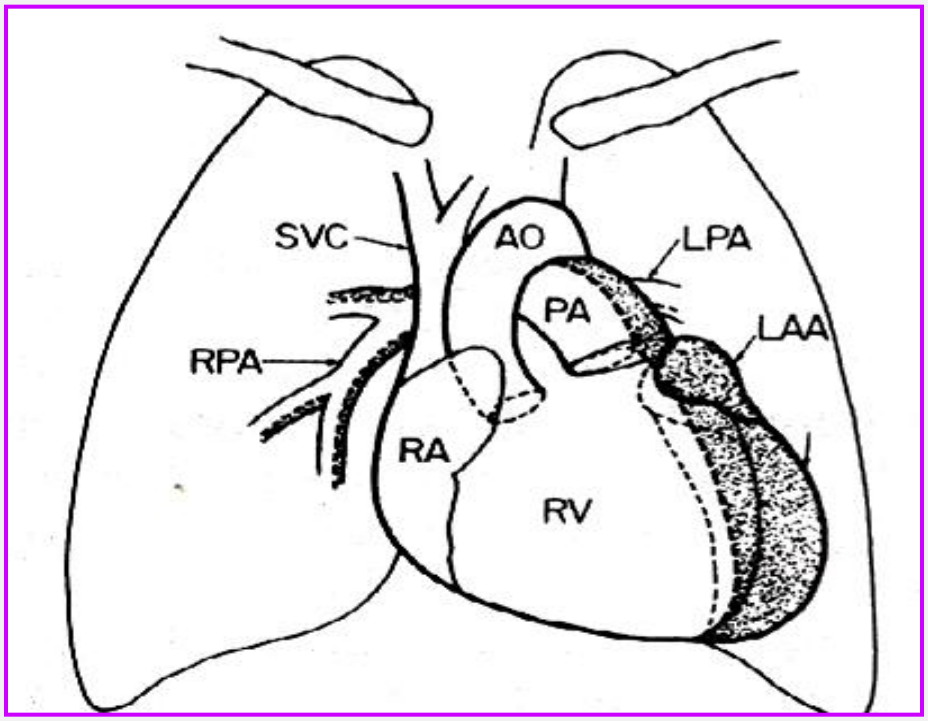




Tele, EKG → **NORMAL**

Orta büyüklükte VSD





Sol kalp boşluklarında hacim yükü

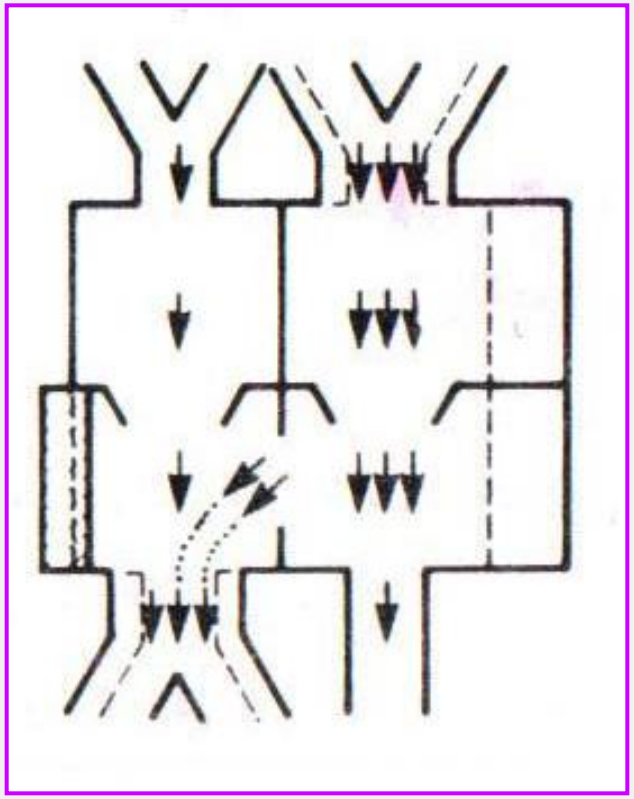
PA basıncı hafif artmış

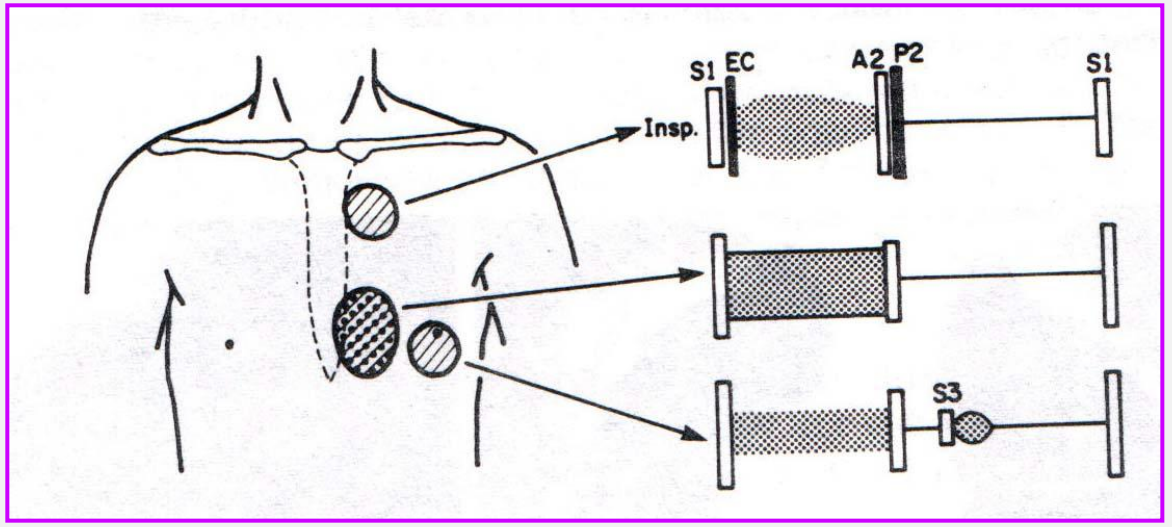
P_2 hafif sert, Pansistolik üfürüm ± thrill, middiyastolik üfürüm

EKG: Sol ventrikül hipertrofisi, sol atriyal genişleme

Tele: Kalp büyük, pulmoner konus belirgin, Akciğer kanlanması artmış

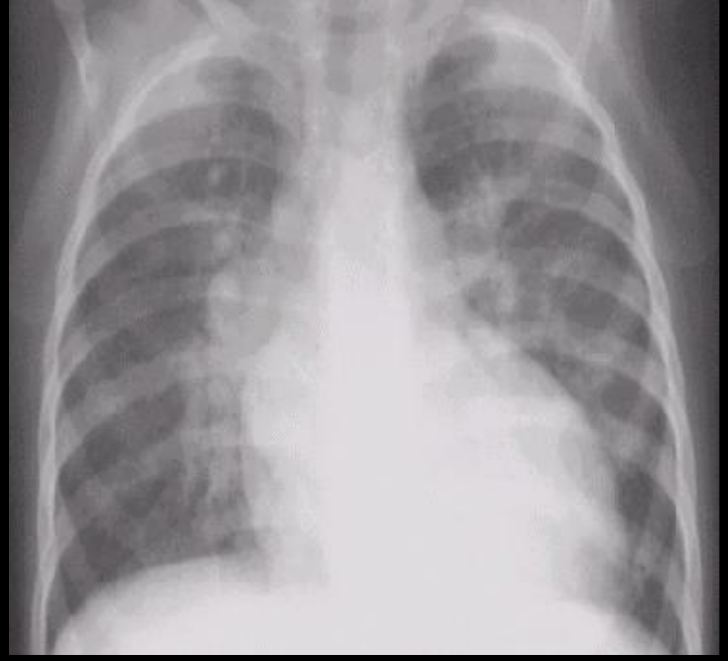
Büyük VSD

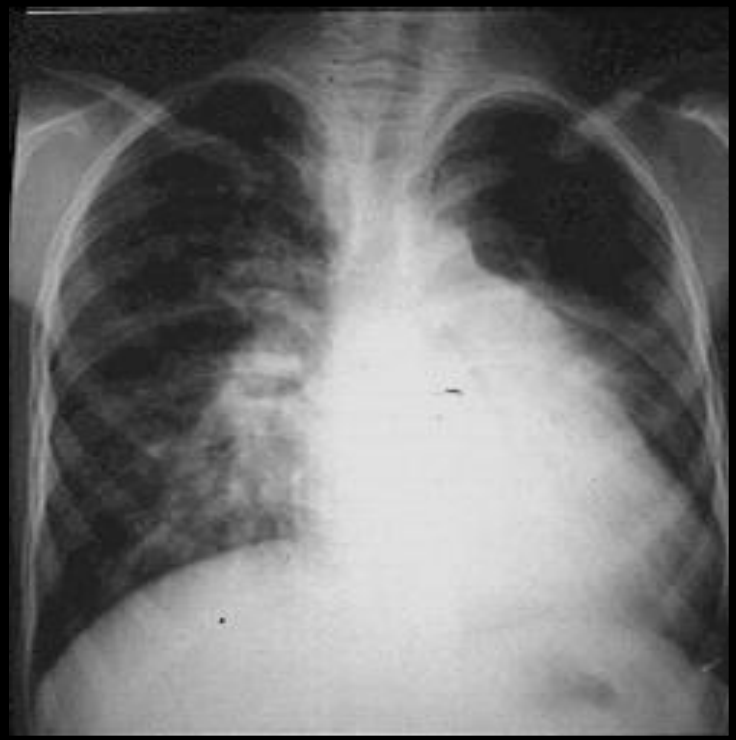


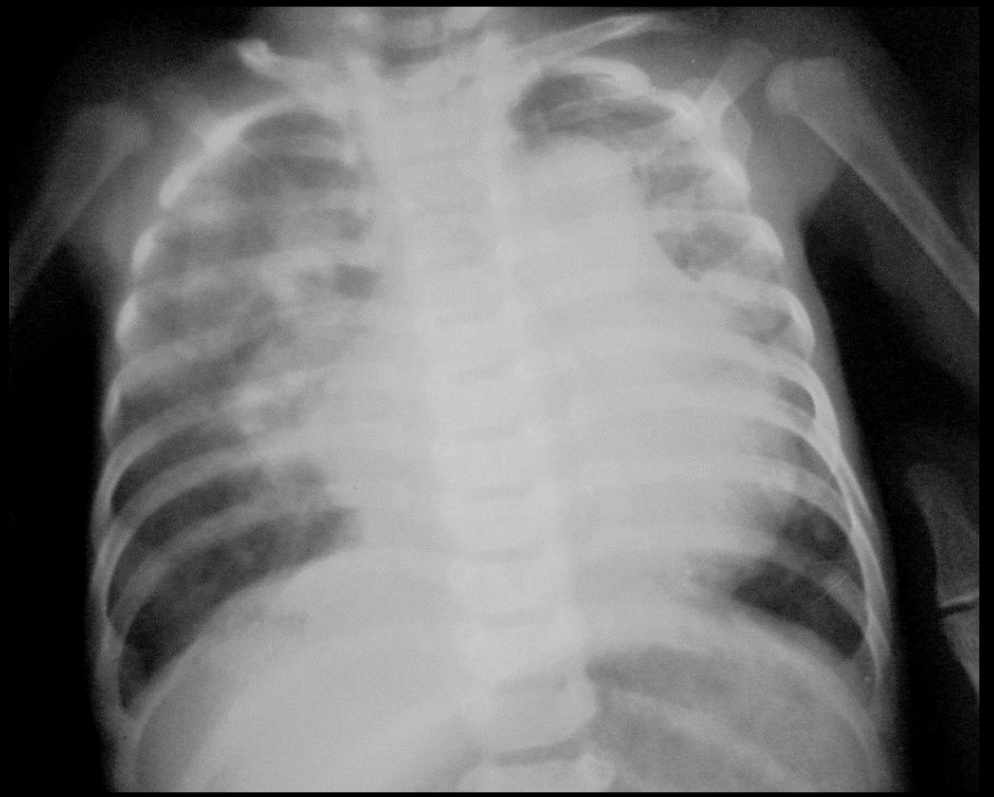


EKG: Biventriküler hipertrofi, sol atriyal genişleme

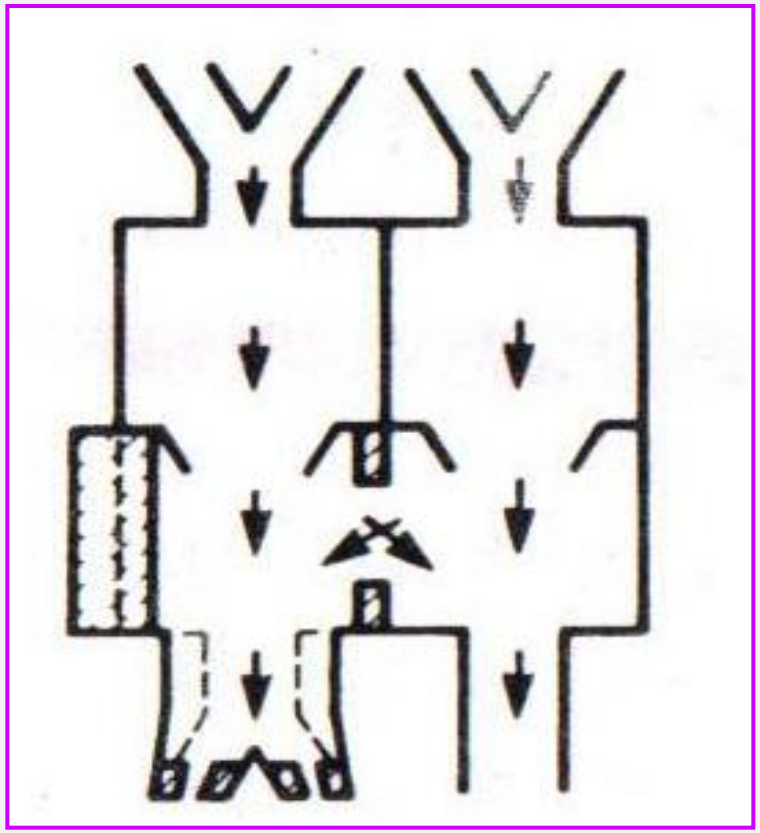
Tele: Kalp büyük, pulmoner konus belirgin, Akc. kanlanması ↑



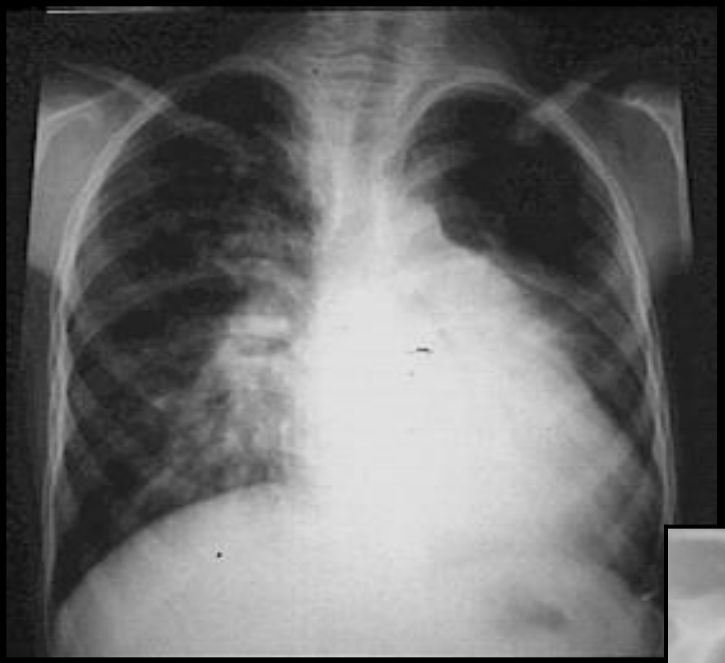


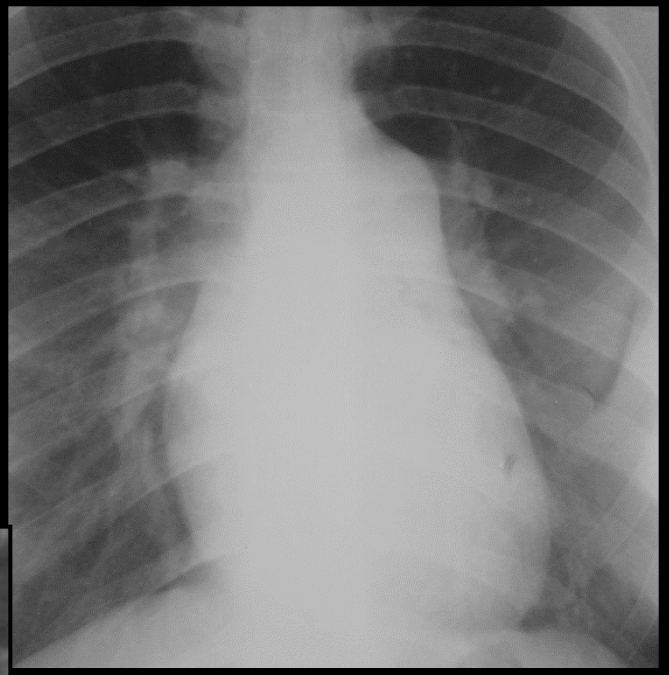


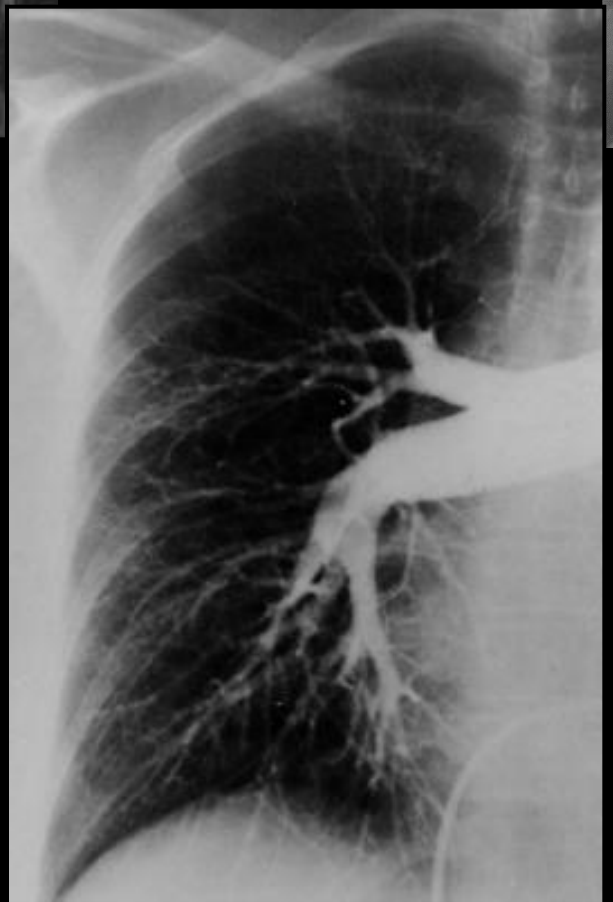
Büyük VSD + Eisenmenger



- Pansistolik üfürüm şiddeti azalır
- Middiyastolik üfürüm kaybolur
- S_2 tek ve sert
- Kalp küçülür
- PA geniş
- Defektten iki yönlü geçiş







ÖYKÜ

Küçük VSD → **Belirtisiz (Asemptomatik)**

Orta-büyük VSD

- **Doğumdan > 4 hft sonra KY bulguları**
- **Sık soluma**
- **Emme güçlüğü**
- **Aşırı terleme**
- **Kilo alamama**
- **Tekrarlayan akciğer enfeks.**

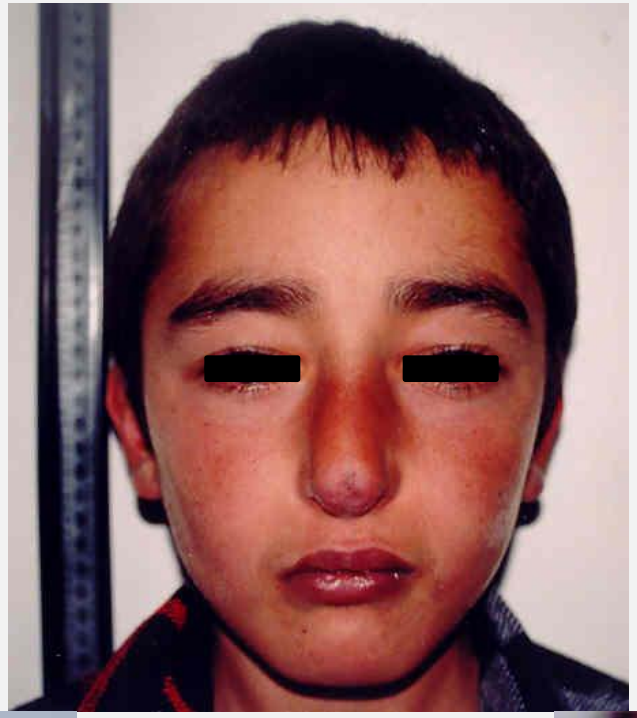
Eisenmenger → **Siyanoz, düşük efor kapasitesi, parmaklarda çomaklaşma**

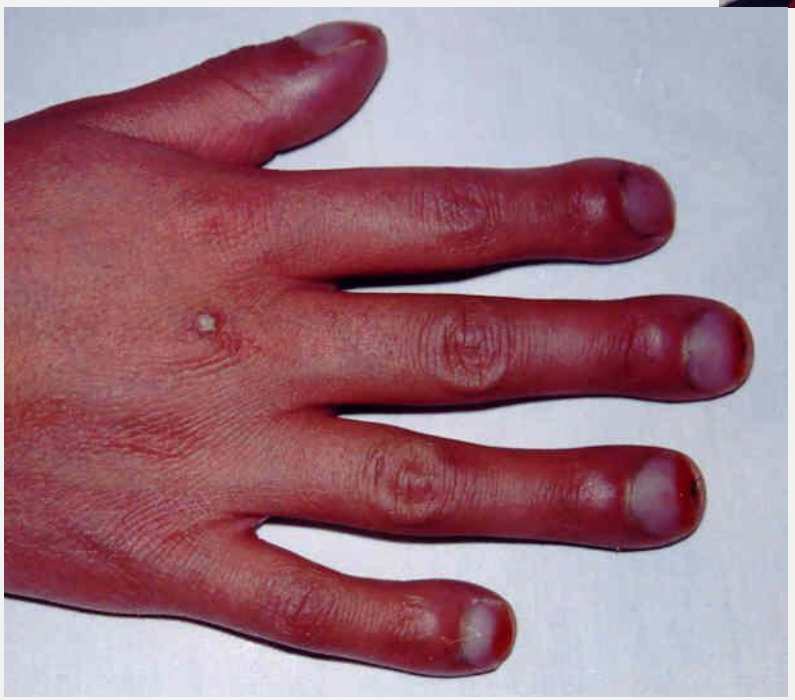
FİZİK İNCELEME

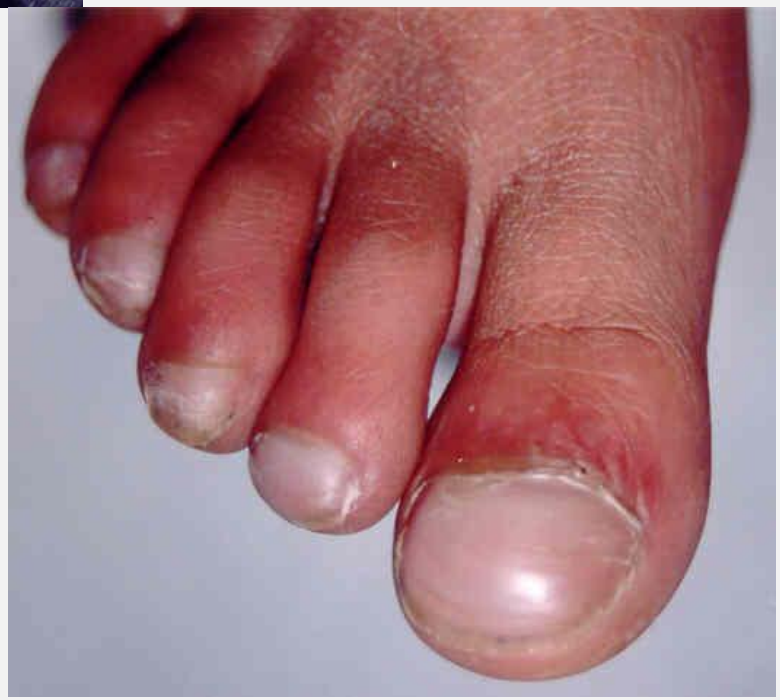
Küçük VSD → **Pansistolik üfürüm ± thrill dışında normal**

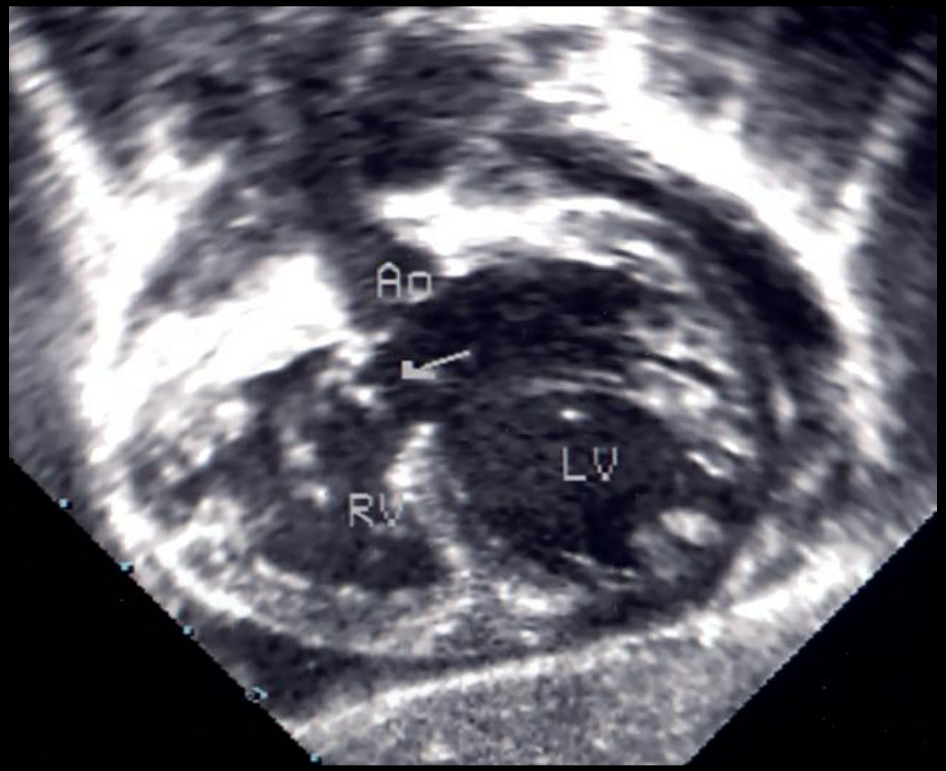
Orta-büyük VSD

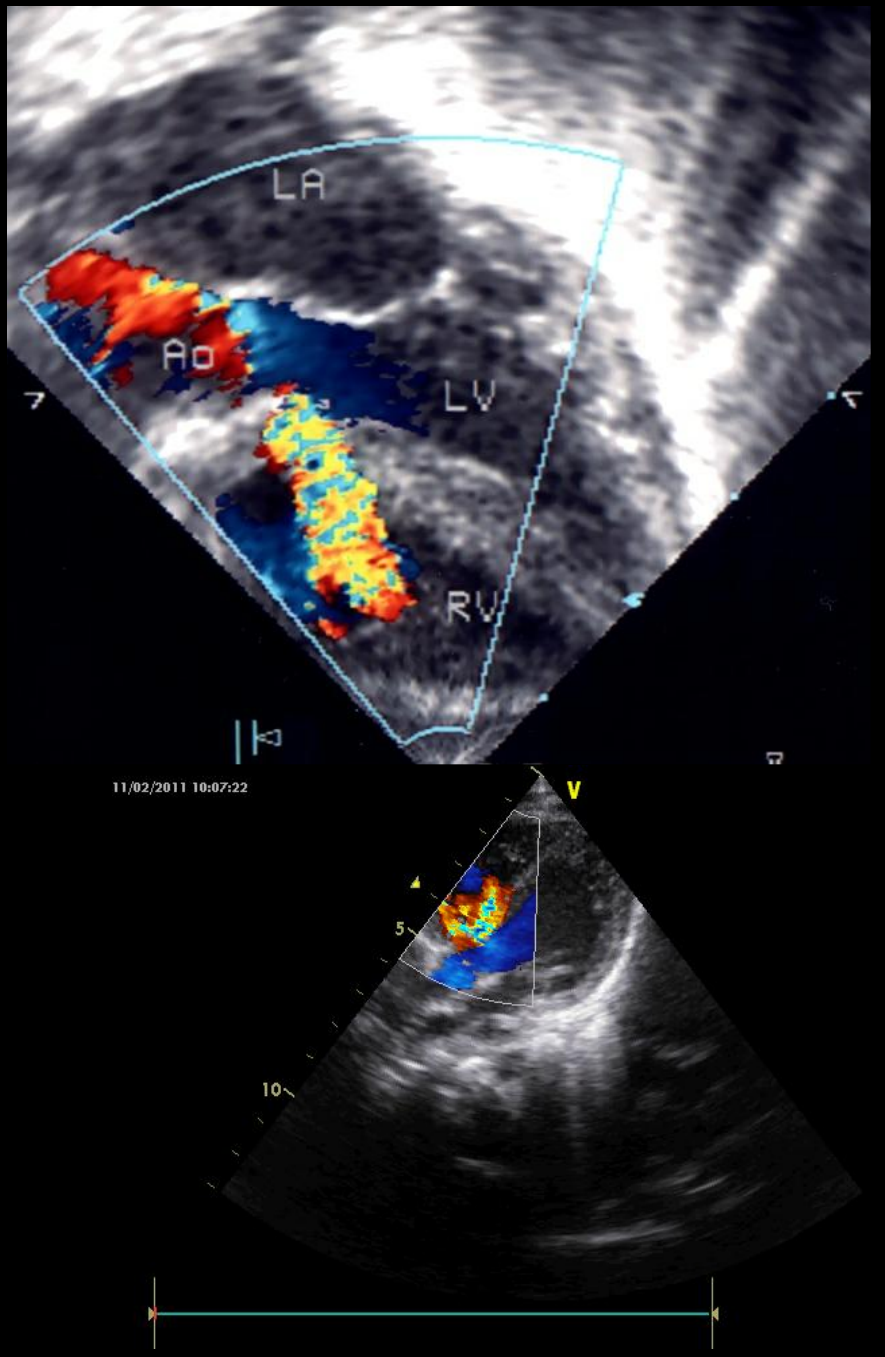
- **Prekordium kabarık**
- **Prekordial aktivite artmış**
- **Parasternal lift**
- **Üfürüm ± thrill**
- **Apikal diyastolik üfürüm**
- **P_2 sert**





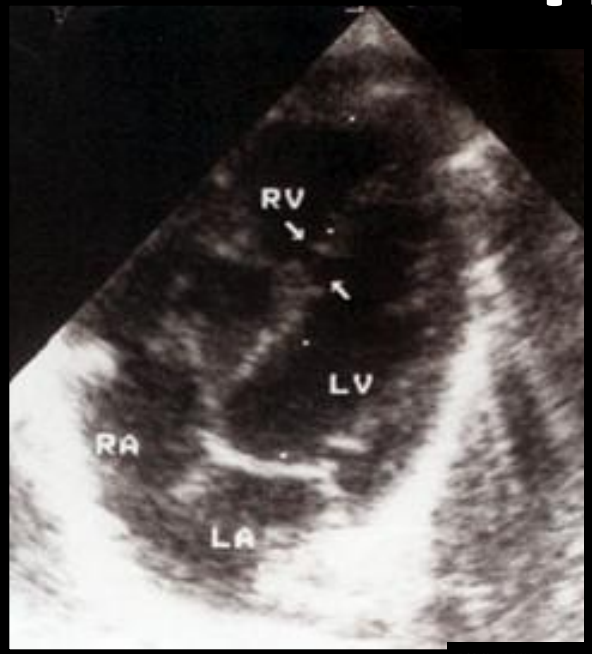


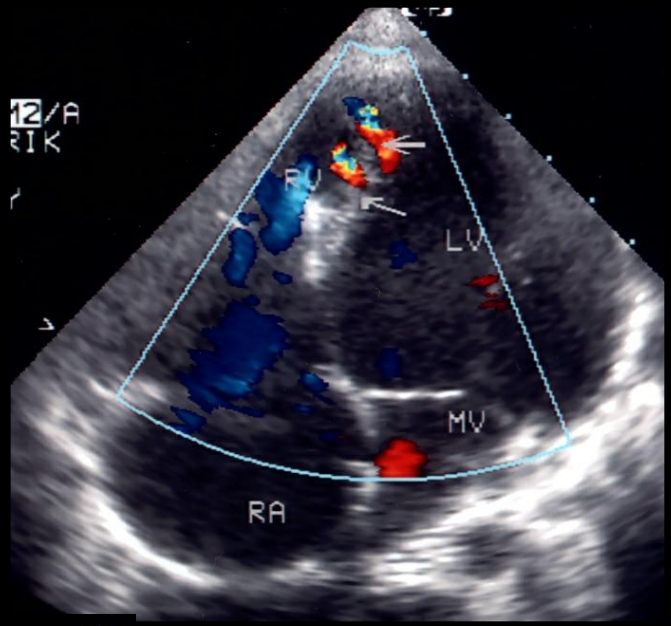


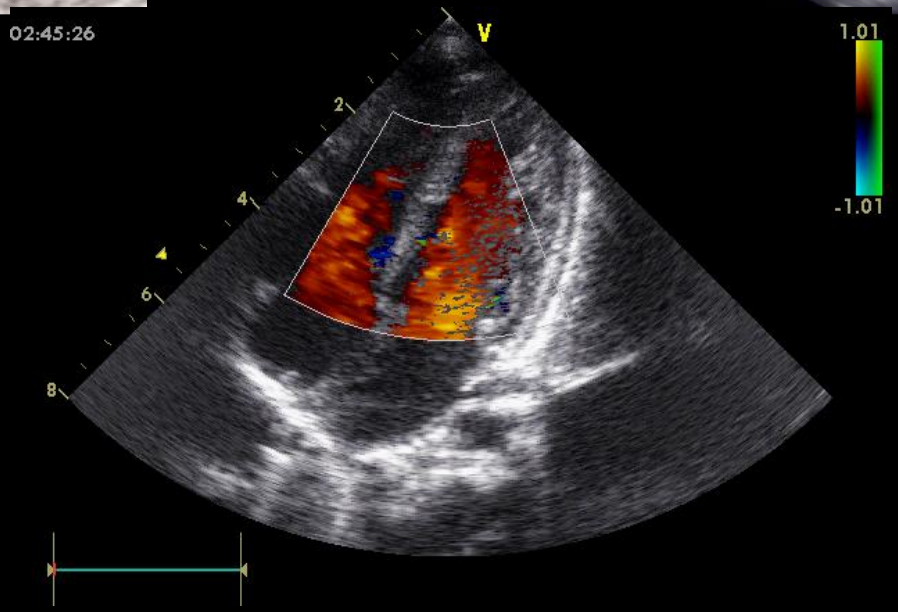


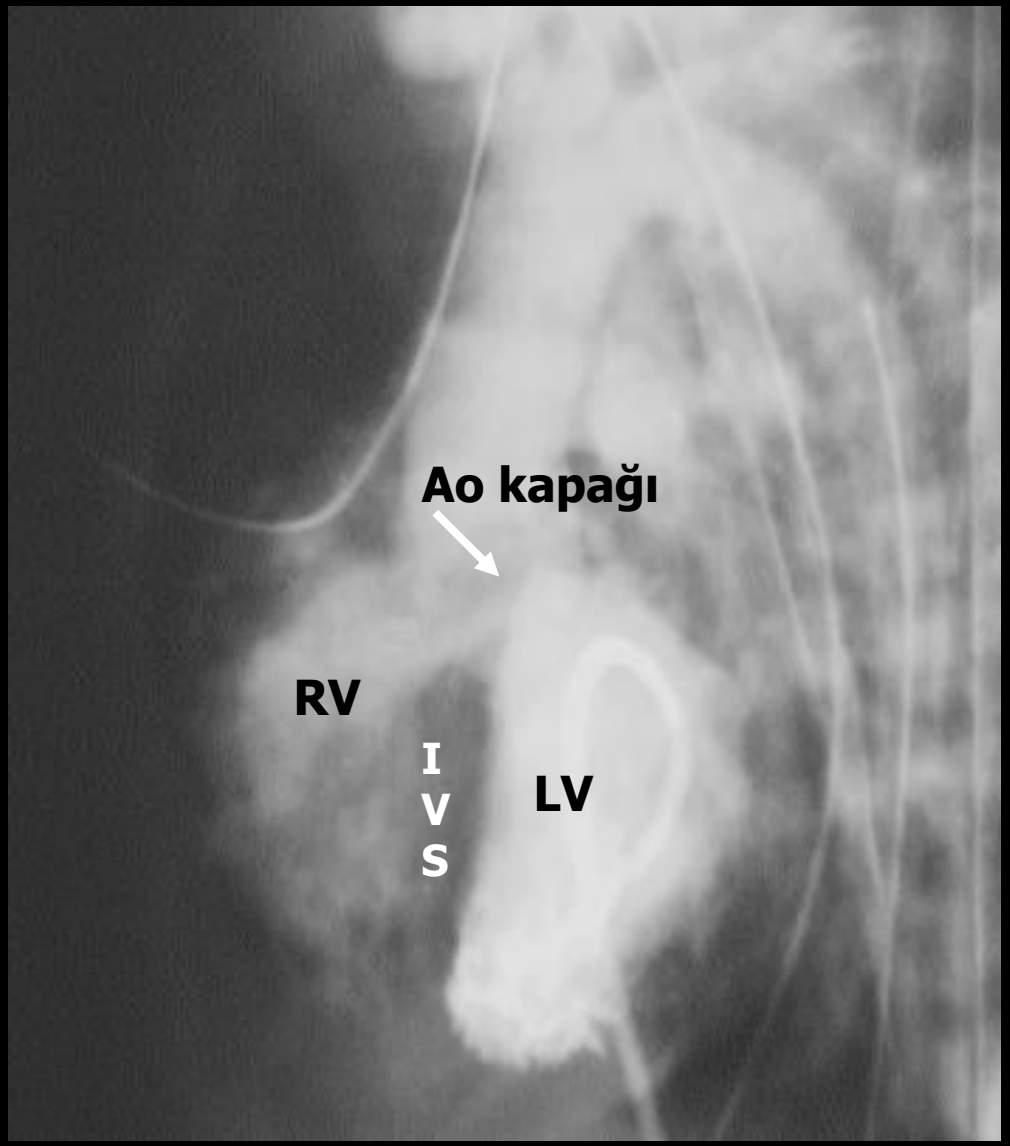
PERİMEMBRANÖZ VSD

TRABEKÜLER VSD





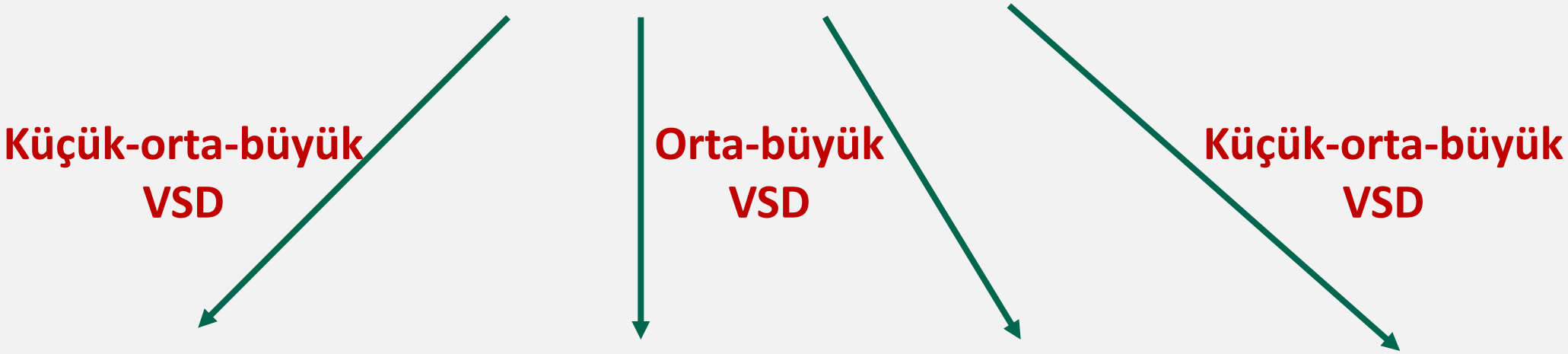




PKA/SKA
PA basıncı
Aort basıncı
PDD
SDD
Ek anomali

Perimembranöz VSD Anjiografi

Gidiş (VSD)



Kendiliğinden küçülme/kapanma

% 30-40
İlk 18-24 ay
Küçük VSD % 70
Trabeküler VSD % 80

Konjestif kalp yetersizliği

> 4-6 hafta

Pulmoner hipertansiyon

Eisenmenger
> 2 yaş

İnfektif endokardit

Ağız hijyeni

Tedavi

- Pulmoner HT yoksa egsersiz kısıtlanmaz
- Ağız hijyeni ve İnfektif endokardit koruması
- Kalp yetersizliği tedavisi
- Beslenmenin düzenlenmesi (Yüksek kalorili)

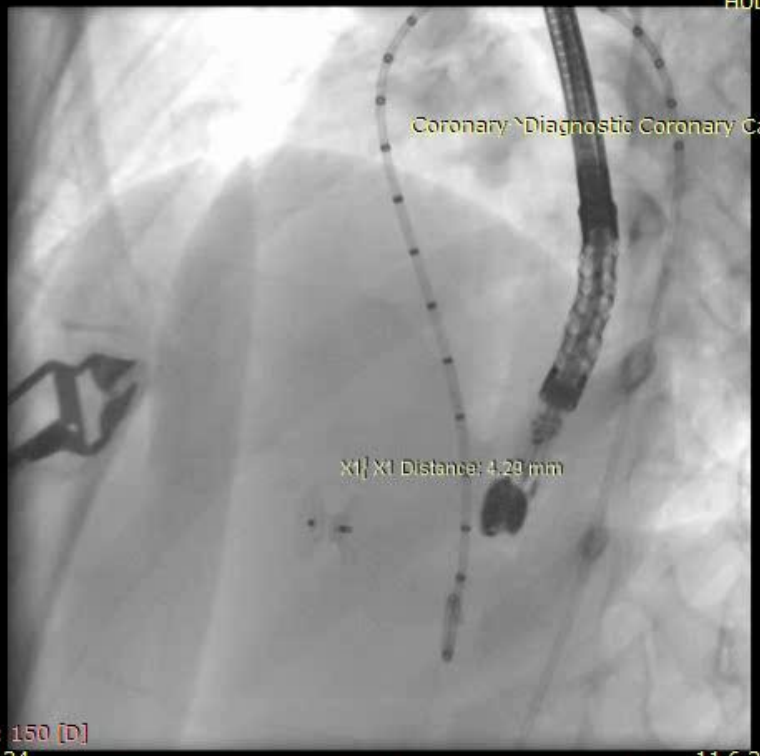
Tedavi

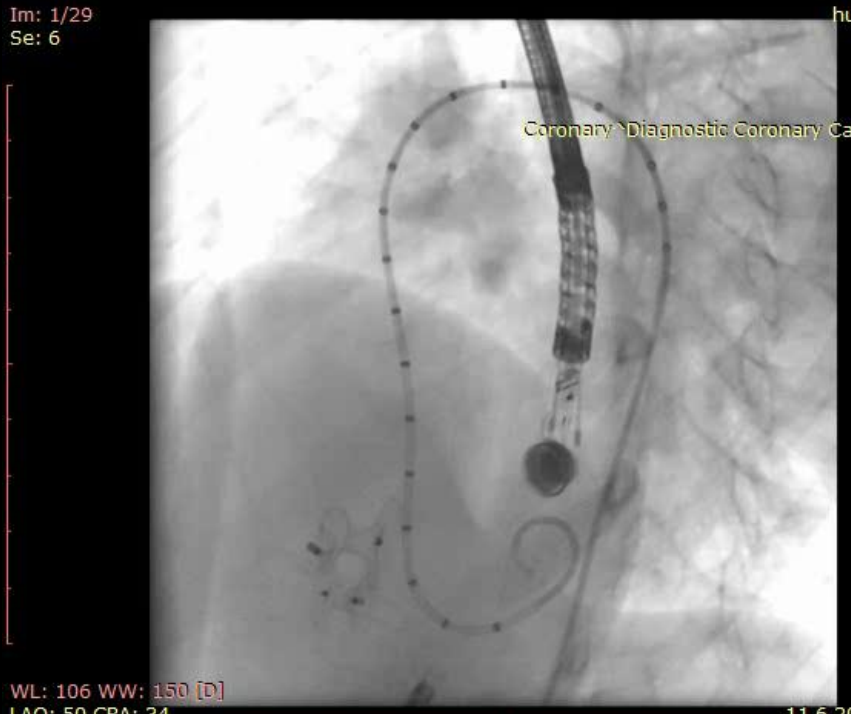
CERRAHİ

< 1 yaş tıbbi tdv yanıtsız KY
>1 yaş sol sağ geçişi fazla (PKA/SKA > 2:1)
PH bulguları (+),KY bulgusu yok: Hemodinamik çalışma

Küçük VSD (PKA/SKA < 1.5) cerrahi yapılmaz

Kateterle kapatma
Cerrahiye bağlı ölüm < % 5
AV tam blok, rezidü şant gibi komplikasyonlar


Im: 1/84
Se: 1
HUDANUR KOPUZ
4246
11.6.2005 F
P
1
Coronary `Diagnostic Coronary Catheterization
Coro LD
X1: X1 Distance: 4.29 mm
WL: 106 WW: 150 [D]
LAO: 50 CRA: 34
11.6.2014 07:54:16


Im: 1/29
Se: 6
hudanur kopuz
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11.6.2005 F
P
1
Coronary `Diagnostic Coronary Catheterization
Coro LD
WL: 106 WW: 150 [D]
LAO: 50 CRA: 34
11.6.2014 09:29:59

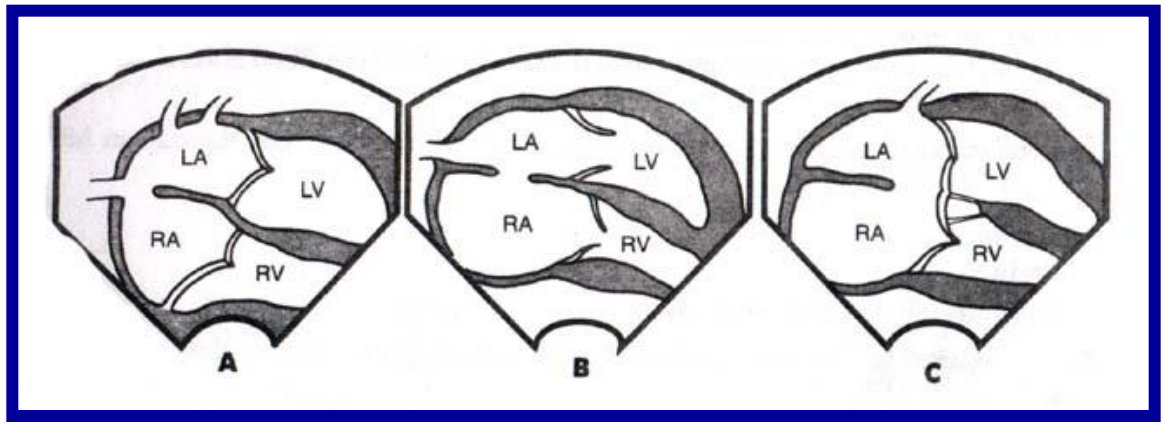
Atriyal Septal Defekt

ASD

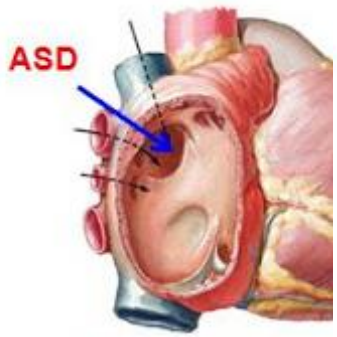
İzole ASD → DKH % 7-10

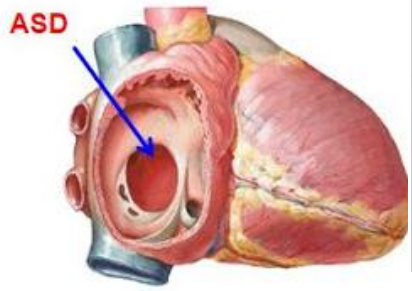
K:E = 2-3/1

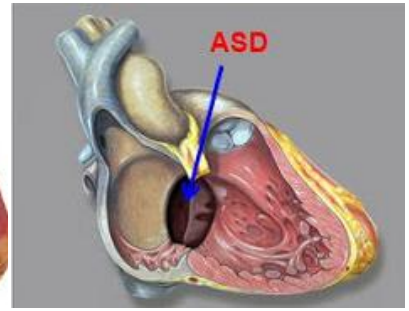
Ailevi ASD (Otoz. dominant)

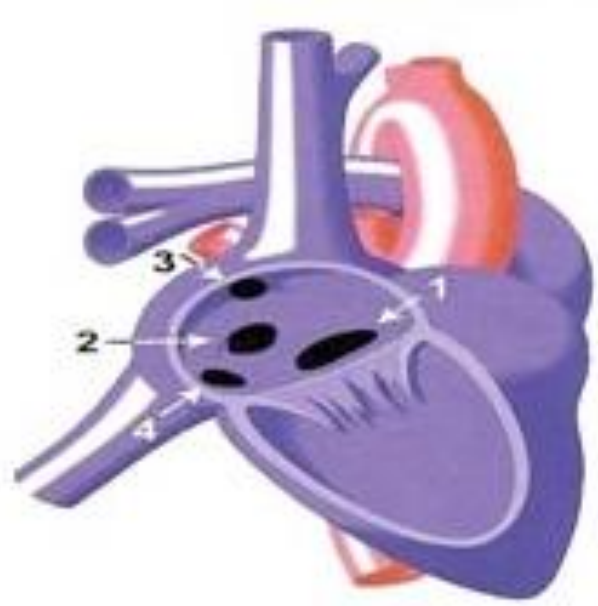


Sinüs venosus	Sekundum	Primum
ASD	ASD	ASD
% 10	% 70-80	% 15







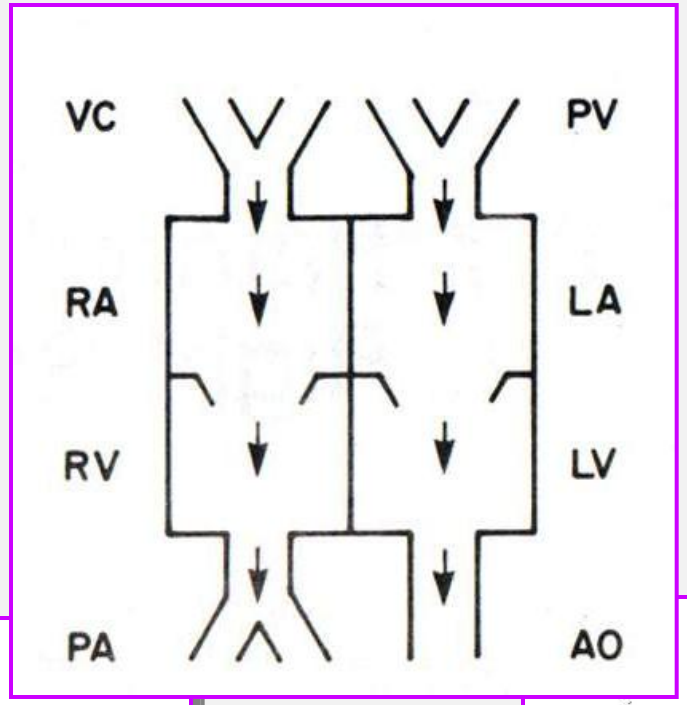


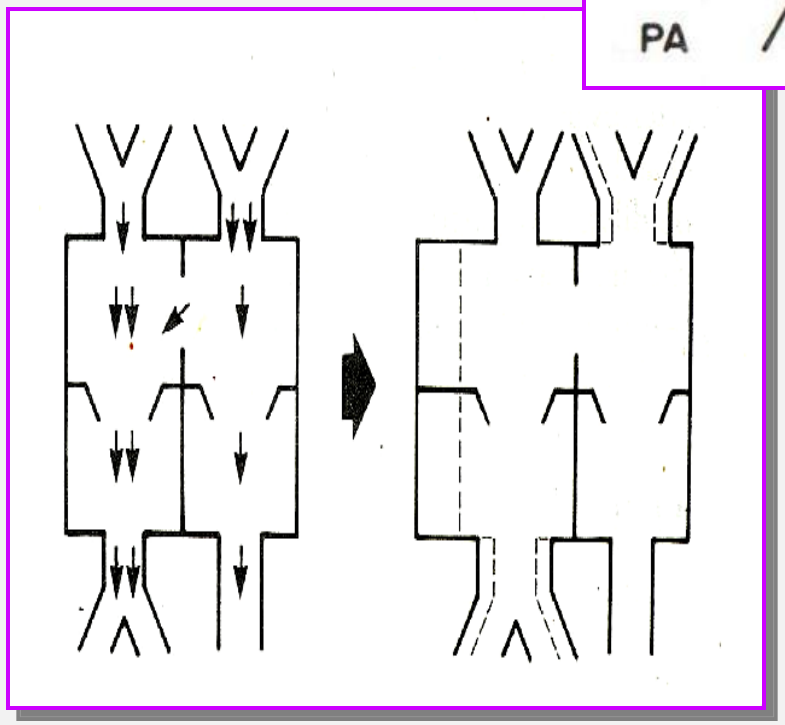
ASD’de geçişin yönünü ve miktarını belirleyen etkenler

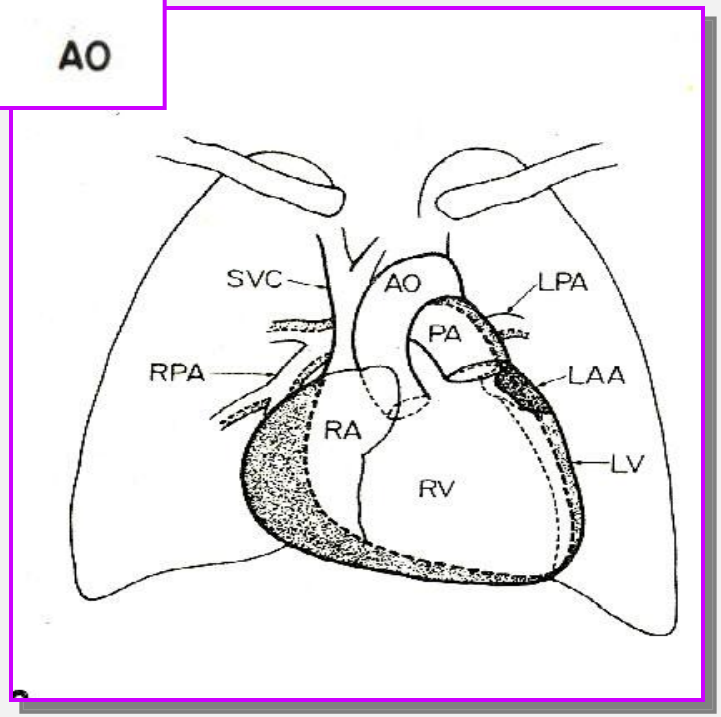
1. **Ventriküllerin genişleyebilirliği (kompliyansı)**
2. **Defektin büyüklüğü**

Sağ ventrikül genişleyebilirliği daha fazla olduğu için geçişin yönü Soldan Sağa

Kalbin sağ yanında diyastolik (hacim) yüklenmeye yol açar.

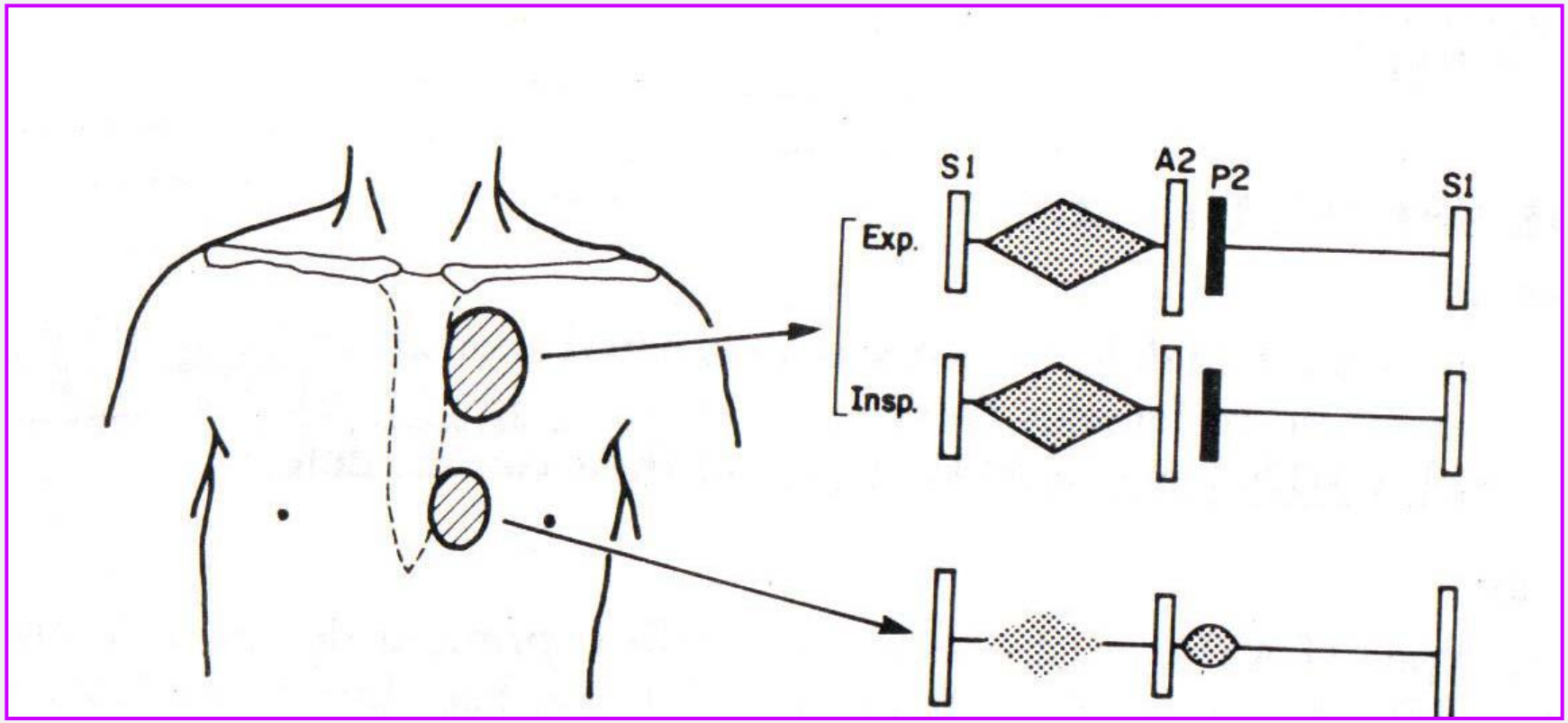

VC
PV
RA
LA
RV
LV
PA
AO

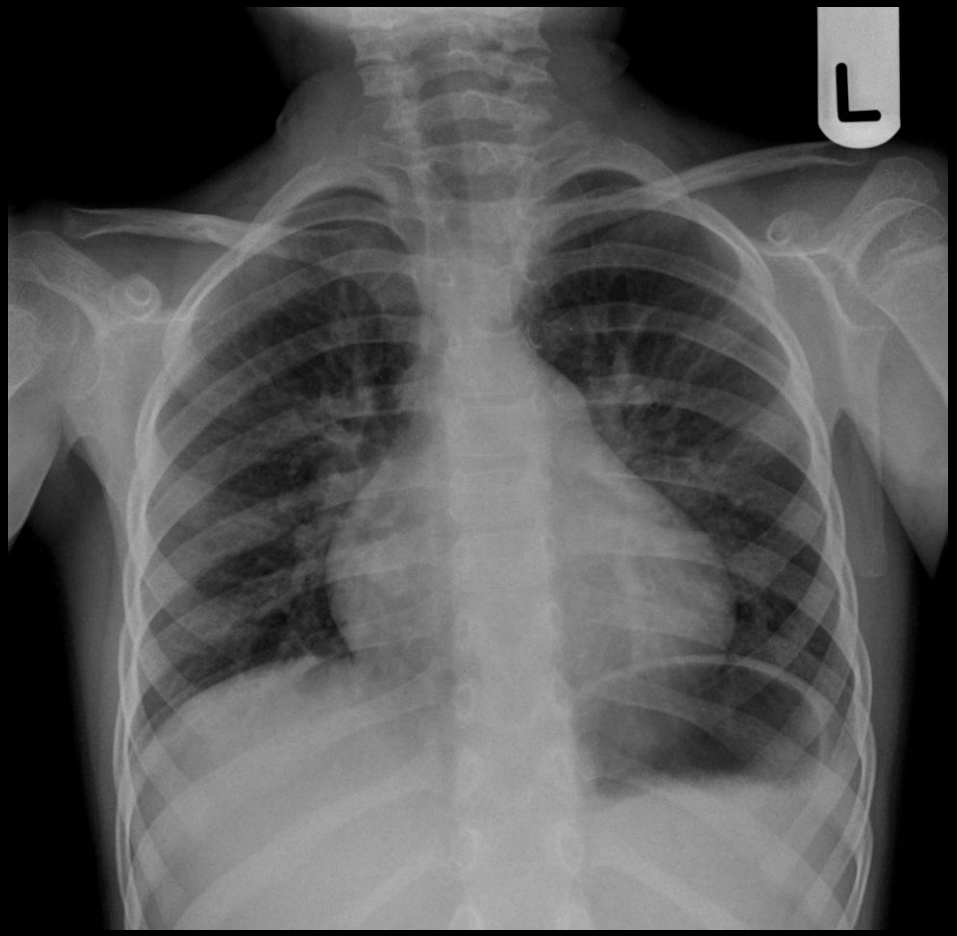



SVC
AO
LPA
PA
RPA
LAA
RA
LV
RV

ASD

Çocuklar genellikle belirtisiz
Nadiren çocuklukta KKY bulguları

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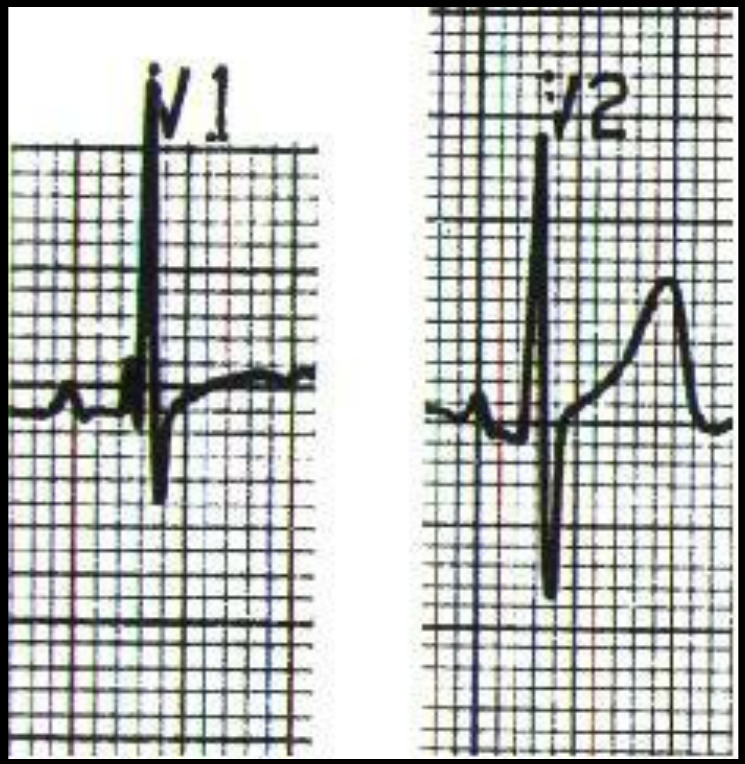
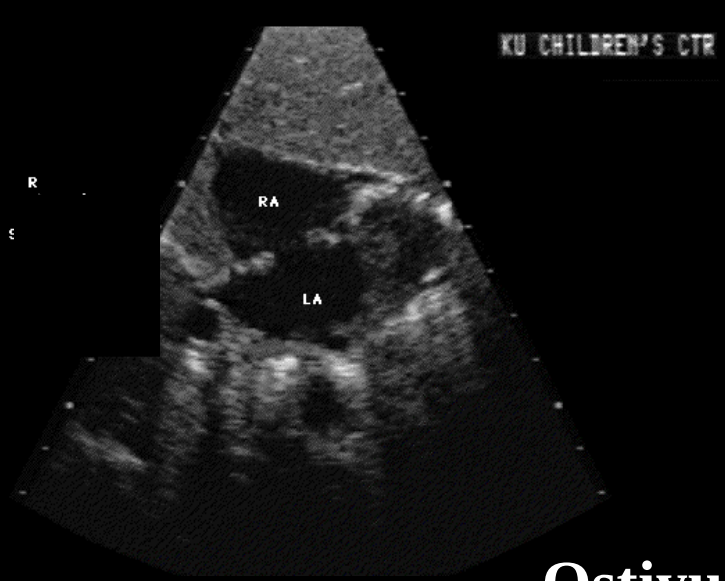
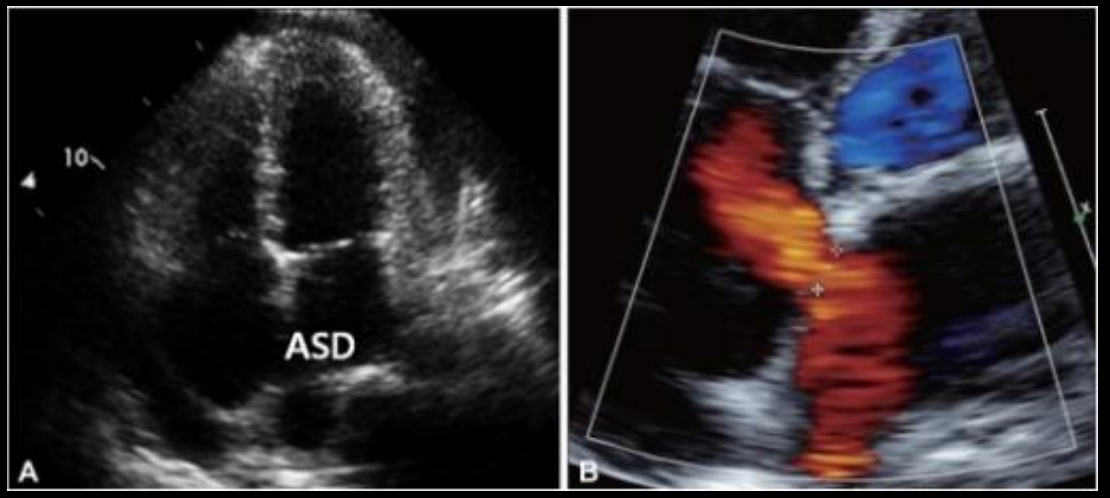
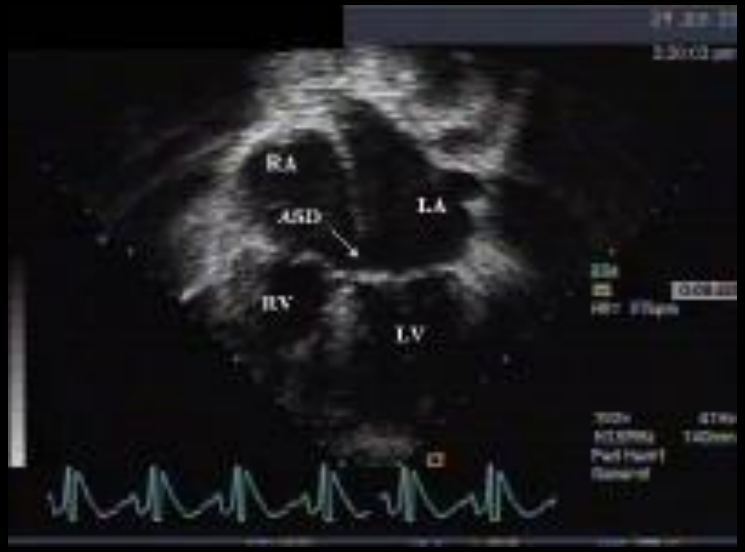

V1
V2



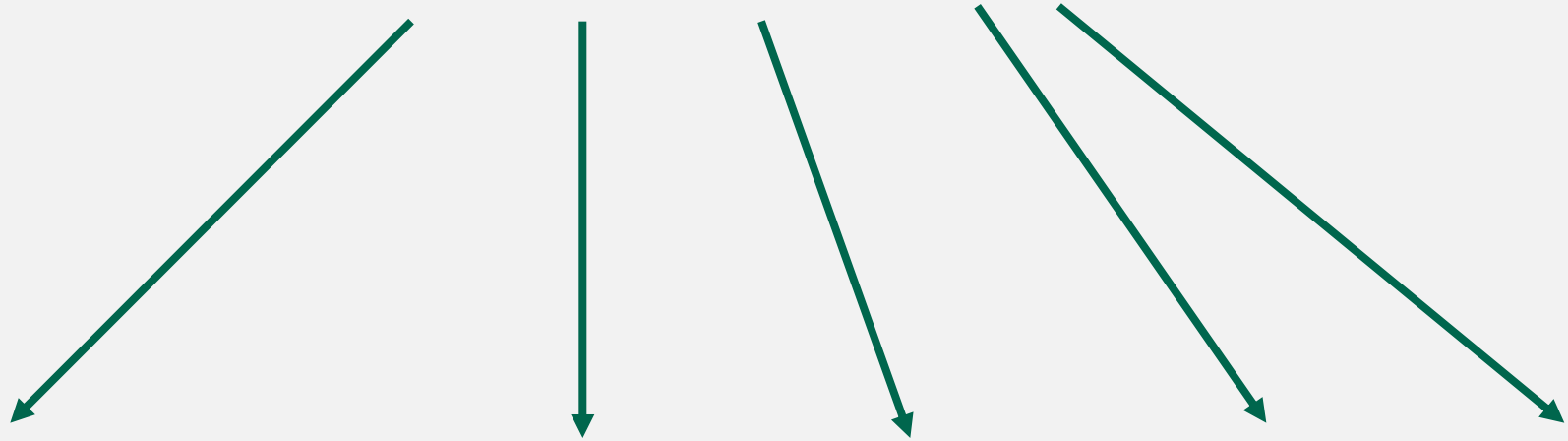


Ostiyum sekundum ASD



Ostiyum Primum ASD

Gidiş (ASD)



Kendiliğinden küçülme/kapanma

Primum ve sinüs venosus küçülüp kapanmaz

< 8mm sekundum ASD ilk 4 yılda % 40 kapanma (foramen ovale ?)

Sağ kalp yetersizliği

20'li yaşlar

Bebeklikte Başka sebep ?

Pulmoner HT

30'lu yaşlar

Atriyal disritmi

A. flutter
A. fibrilasyon

Paradoks emboli

İnfektif endokardit

Yalnızca ostiyum primum ASD

Tedavi (ASD)

- Egsersiz kısıtlanmaz
- İnfektif endokardit koruması gereksiz
  - Primum ASD hariç

Küçük ASD kapatılmaz
Sağ kalp boşlukları geniş değil
PKA/SKA < 1.5

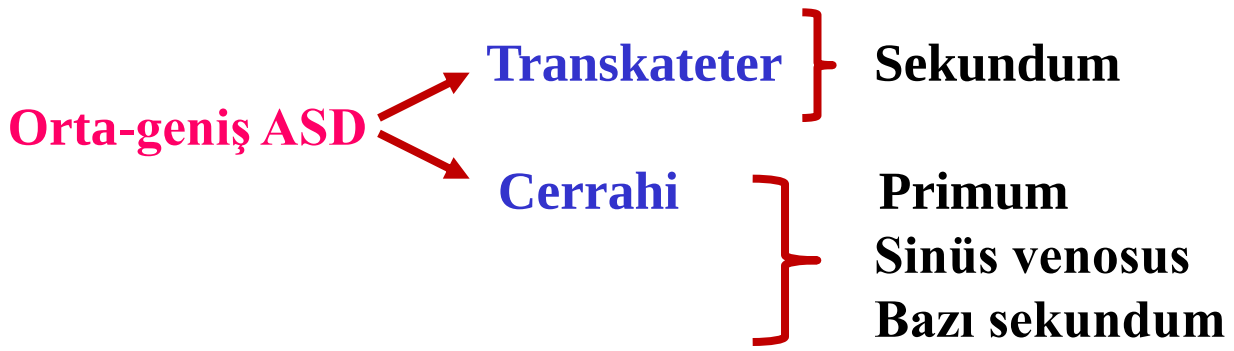
Ölüm < % 1

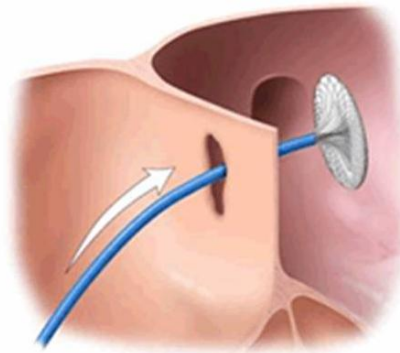
Ameliyat sonrası komplikasyon

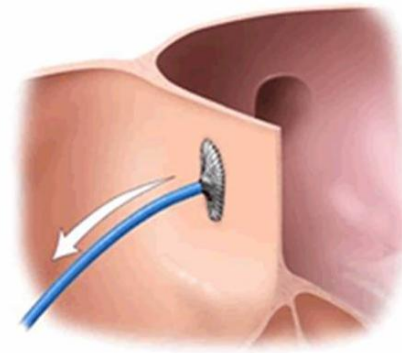
Hasta sinüs sendromu

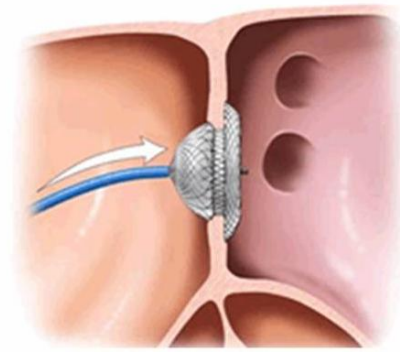
Atriyal disritmiler

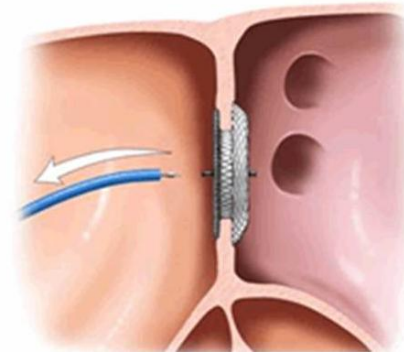
% 20











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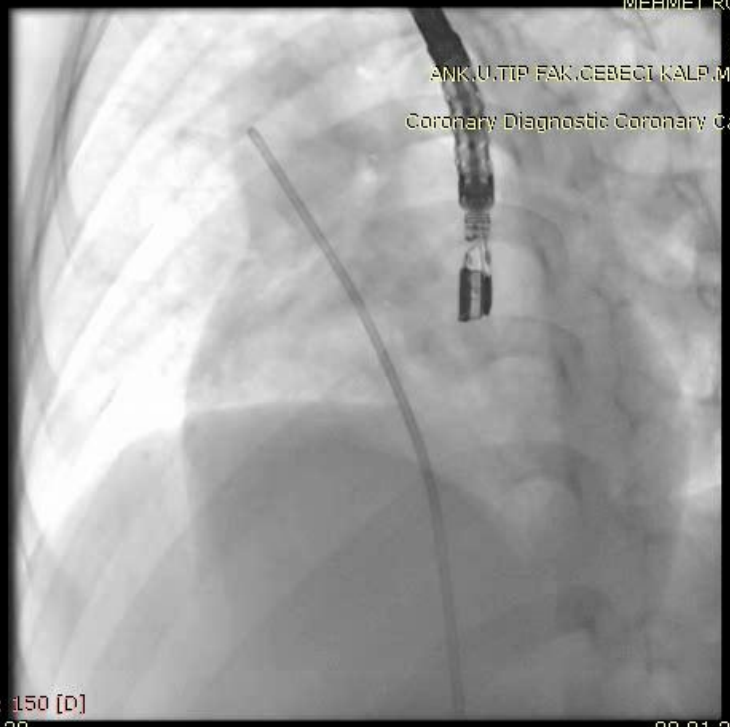
MEHMET RONAY SUNGUR
3882
09.01.2005 M
ANK.U.TIP FAK.CEBECI KALP.MRK//3D735D//
1
Coronary Diagnostic Coronary Catheterization
Coro LD



WL: 106 WW: 150 [D]
LAO: 31 CRA: 29
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Im: 1/130
Se: 3

MEHMET RONAY SUNGUR
3882
09.01.2005 M
ANK.U.TIP FAK.CEBECI KALP.MRK//3D735D//
1
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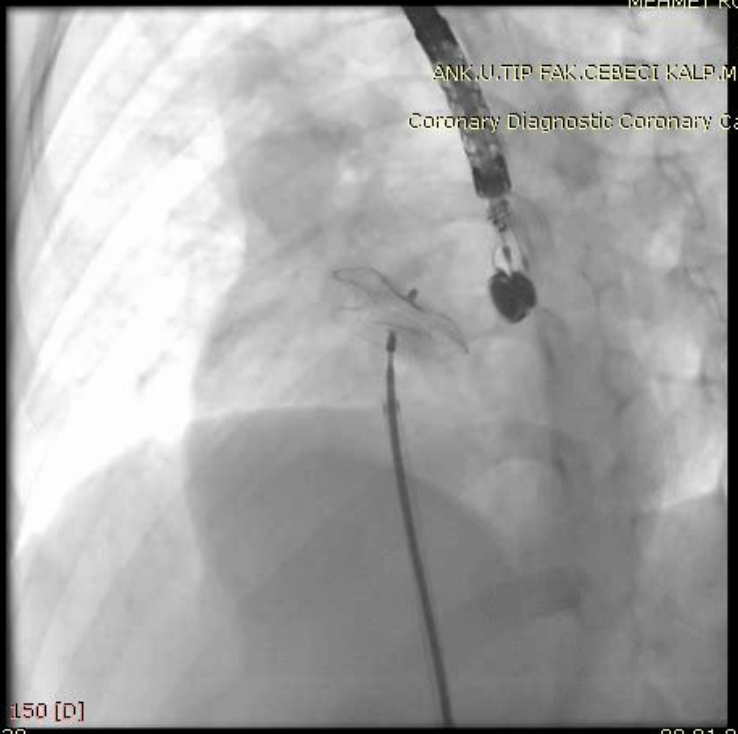
Coro LD



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LAO: 30 CRA: 30
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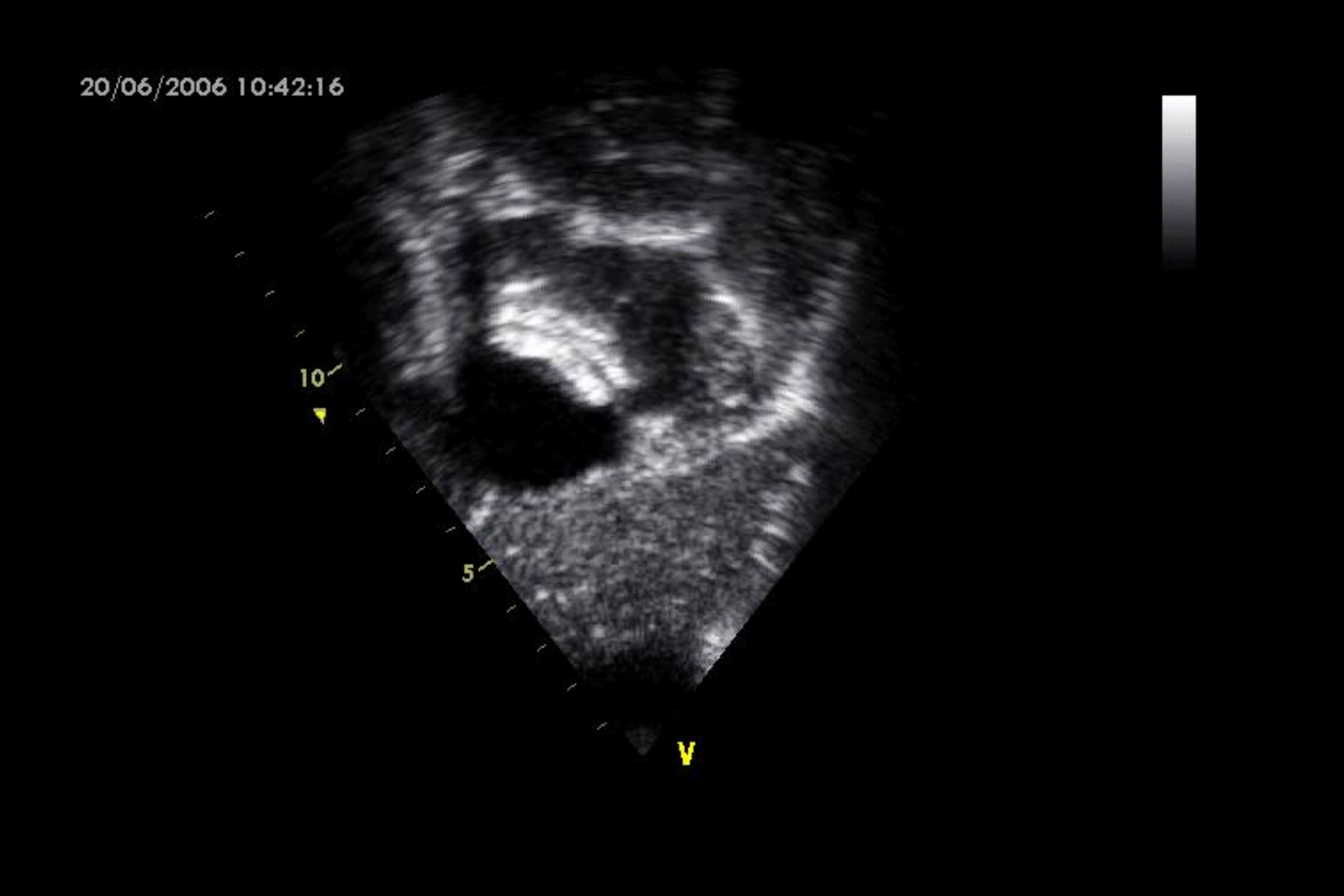
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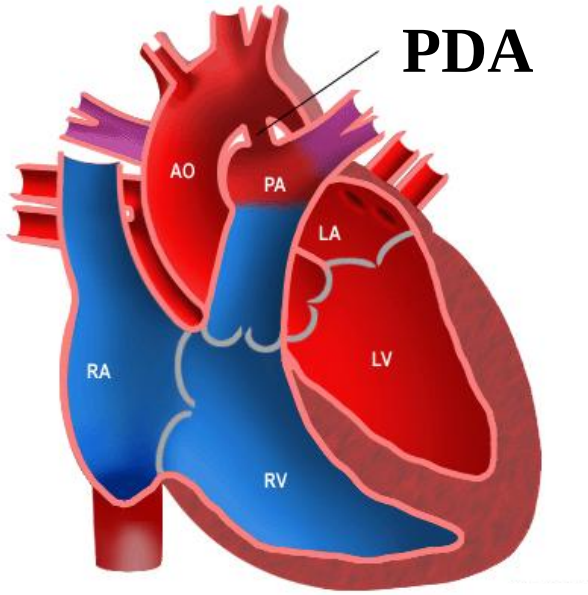
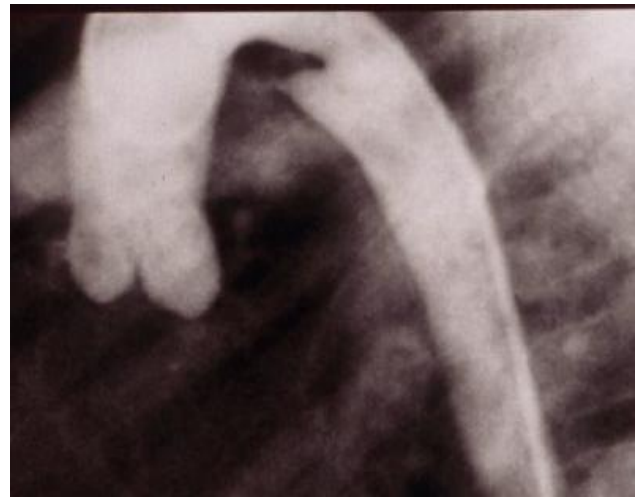
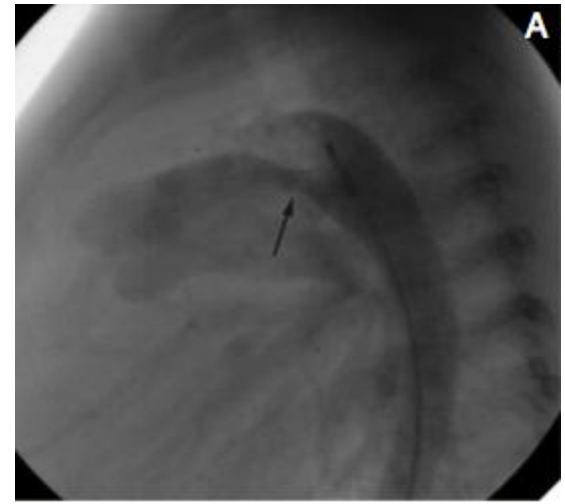
Patent Duktus Arteriyosus







% 5-10 DKH (Preterm PDA hariç)

Pretermler: < 1750 gr % 45

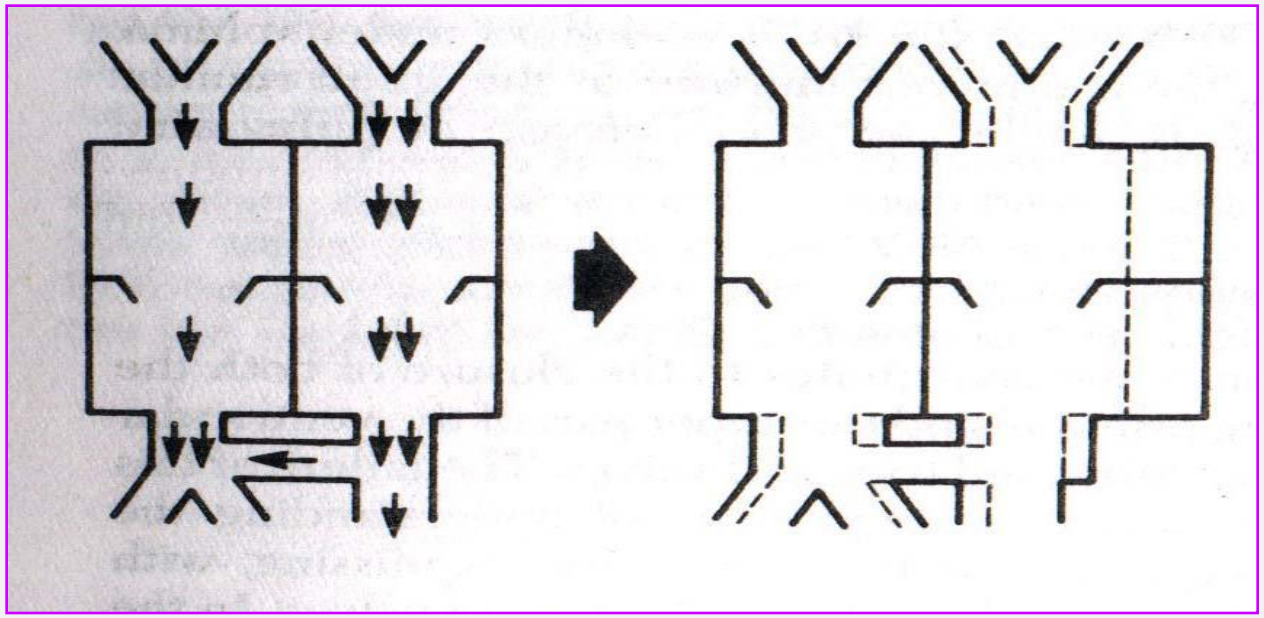
< 1200 gr % 75

Konjenital rubella

Hemodinami VSD'ye benzer

Şantın miktarını belirleyen etkenler

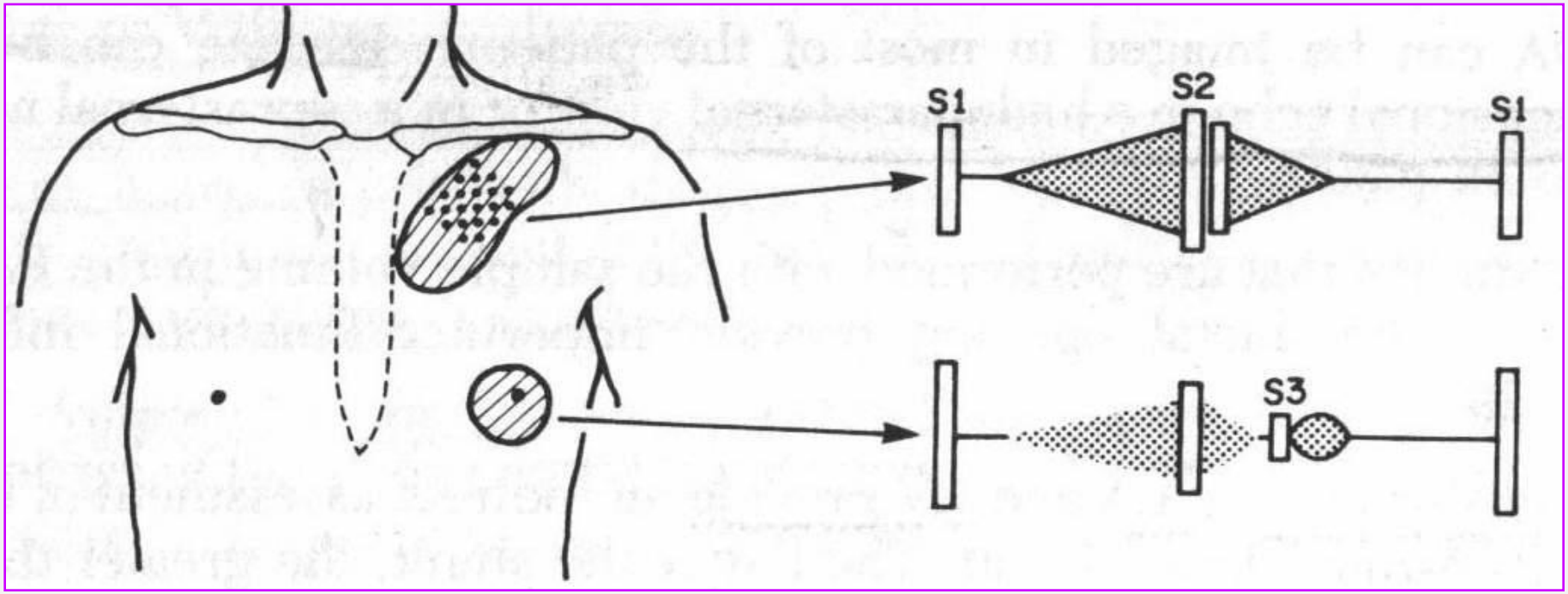
1. **Duktusun büyüklüğü, uzunluğu, kıvrımlı**
2. **Pulmoner damar direncinin derecesi**



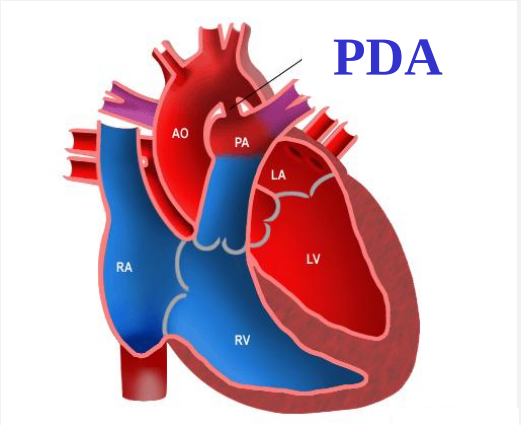
Preterm bebekler dışında

4 haftadan önce KY bulgusu yok

Fizik İnceleme (PDA)



* Prekordium hiperaktif
* Nabızlar sıçrayıcı
* Diferansiyel siyanoz
* PH varsa P_2 şiddetli
* Takikardi, takipne



PDA

Tele: Kalp büyük
Pulmoner konus belirgin
Akciğer kanlanması artmış
EKG: Sol ventrikül hipertrofisi
Biventriküler hipertrofi
Sağ ventrikül hipertrofisi
EKO: Duktusun büyüklüğü
Şantın yönü

Gidiş (PDA)

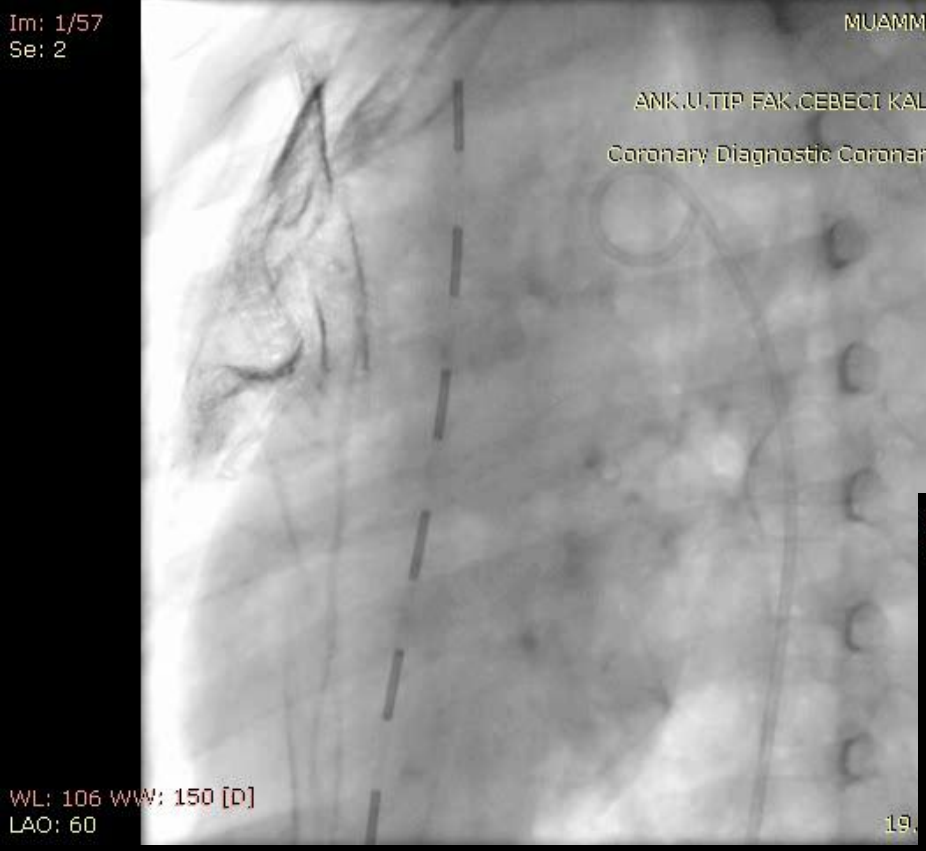
- Pretermler kendiliğinden kapanma
- Kalp yetersizliği
- Tekrarlayan Akc enfeksiyonları
- Pulmoner HT, Eisenmenger
- İnfektif endarterit

Tedavi (PDA)

PRETERM PDA: Sıvı kısıtlaması
Diüretik
İndomethacin, İbuprofen, Paracetamol

- Pulmoner HT yoksa egsersiz kısıtlanmaz
- Ağız hijyeni ve İnfektif endarterit koruması
- Kalp yetersizliği tedavisi
- Beslenmenin düzenlenmesi

- Girişimsel kateter yöntemleri ile kapatma
  Şemsiye, coil vb
- Cerrahi kapatma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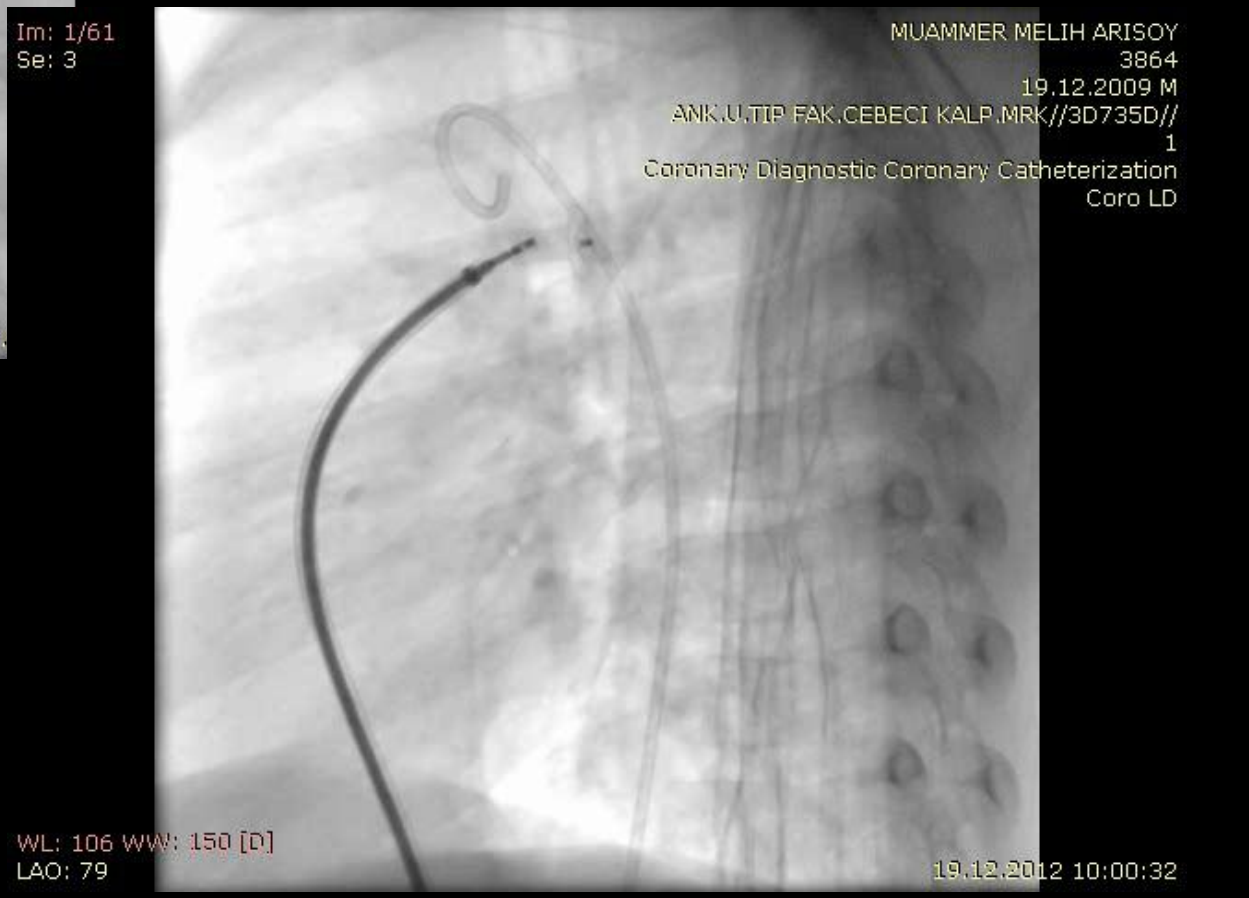
MUAMMER MELIH ARISOY
3864
19.12.2009 M
ANK.U.TIP FAK.CEBECI KALP.MRK//3D735D//
1
Coronary Diagnostic Coronary Catheterization
Coro LD
WL: 106 WW: 150 [D]
LAO: 60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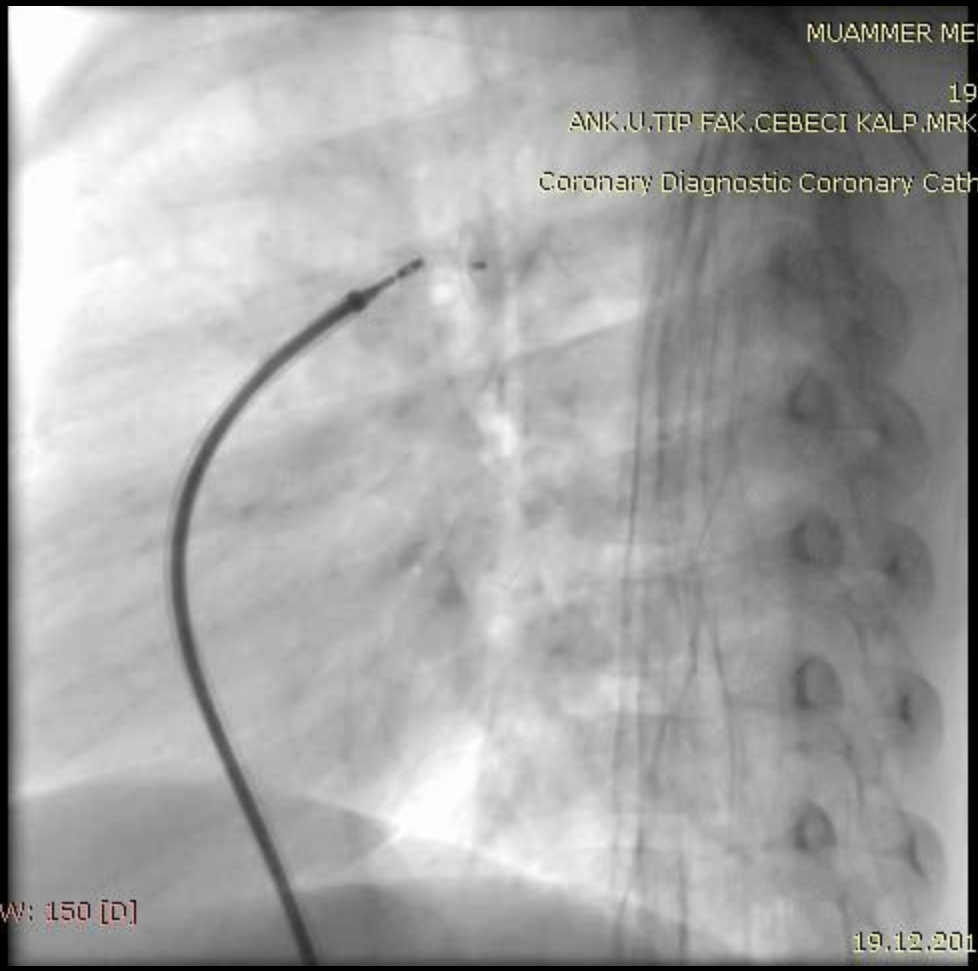
Coro LD
WL: 106 WW: 150 [D]
LAO: 79
19.12.2012 10:00:32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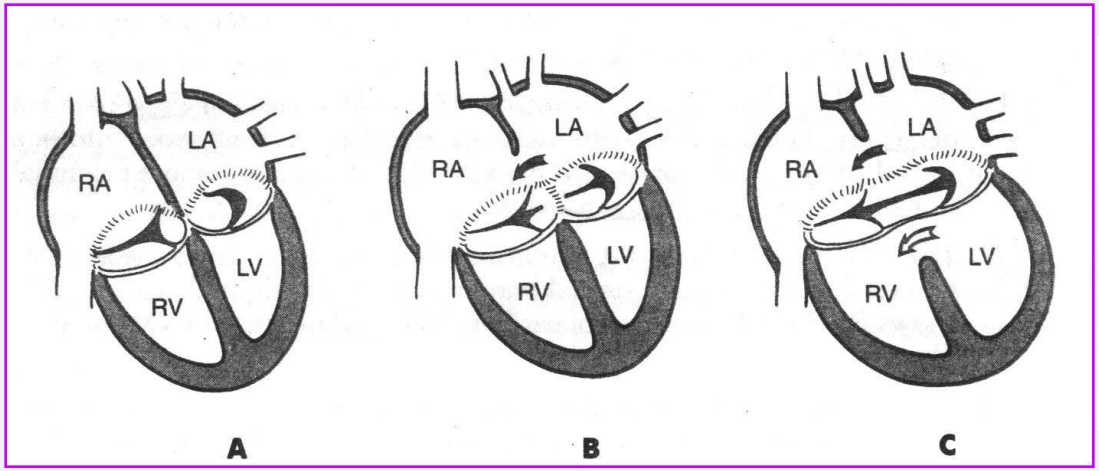
1
Coronary Diagnostic Coronary Catheterization
Coro LD
WL: 106 WW: 150 [D]
LAO: 79
19.12.2012 10:00:42

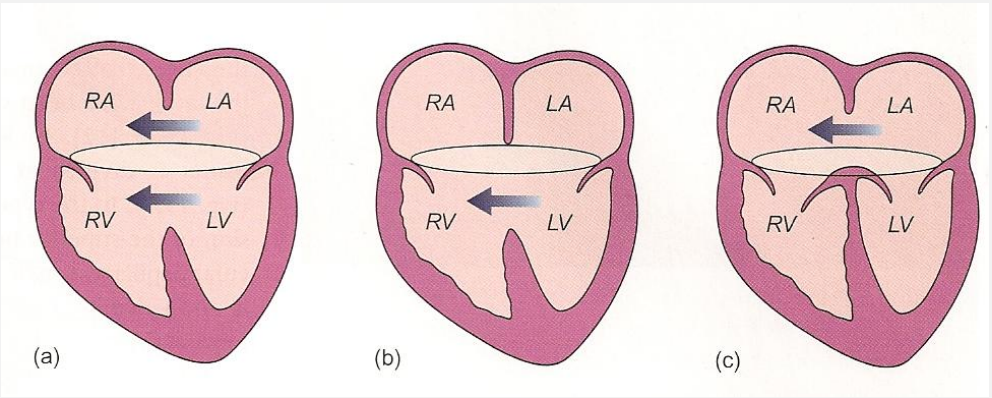
Atriyoventriküler Septal Defekt

AVSD

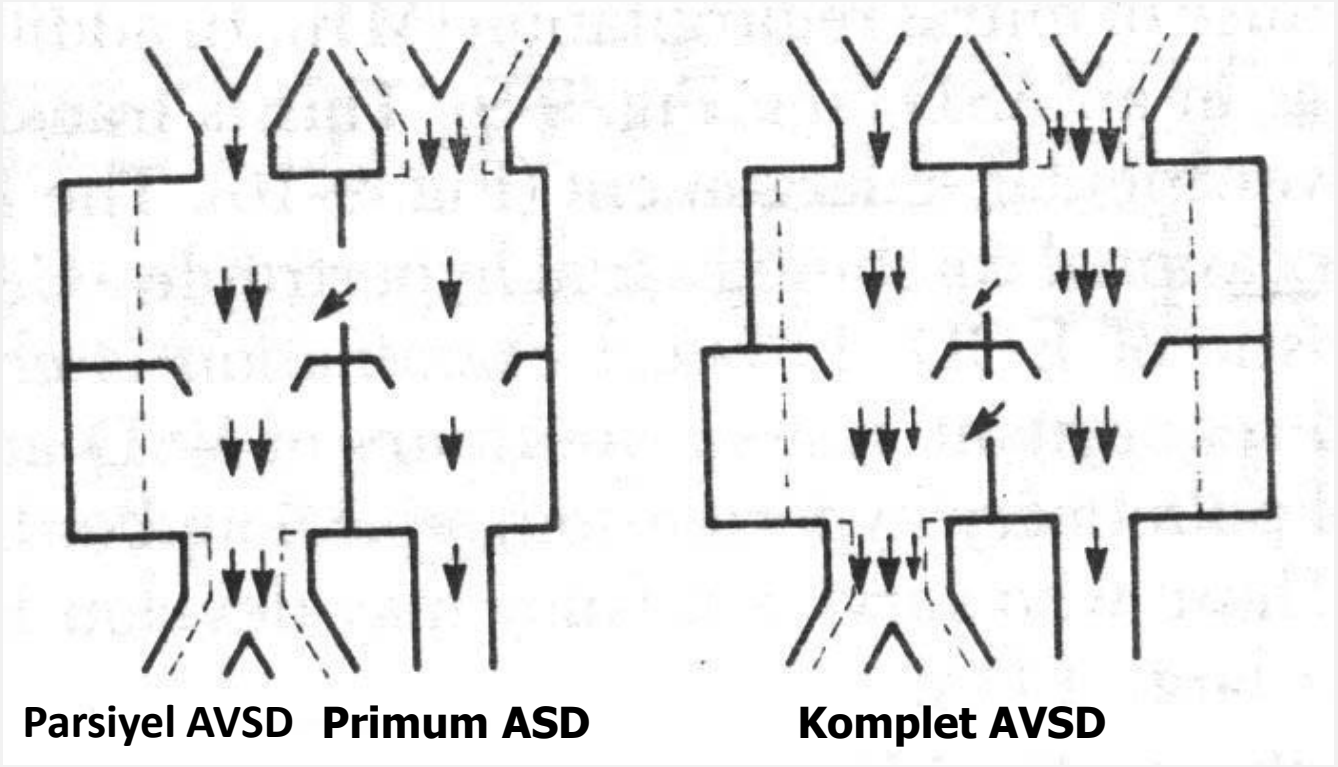
- ✓ Endokardiyal yastık defekti
- ✓ AV kanal defekti

Komplet AVSD → DKH % 2





AVSD



Parsiyel AVSD Primum ASD

Komplet AVSD

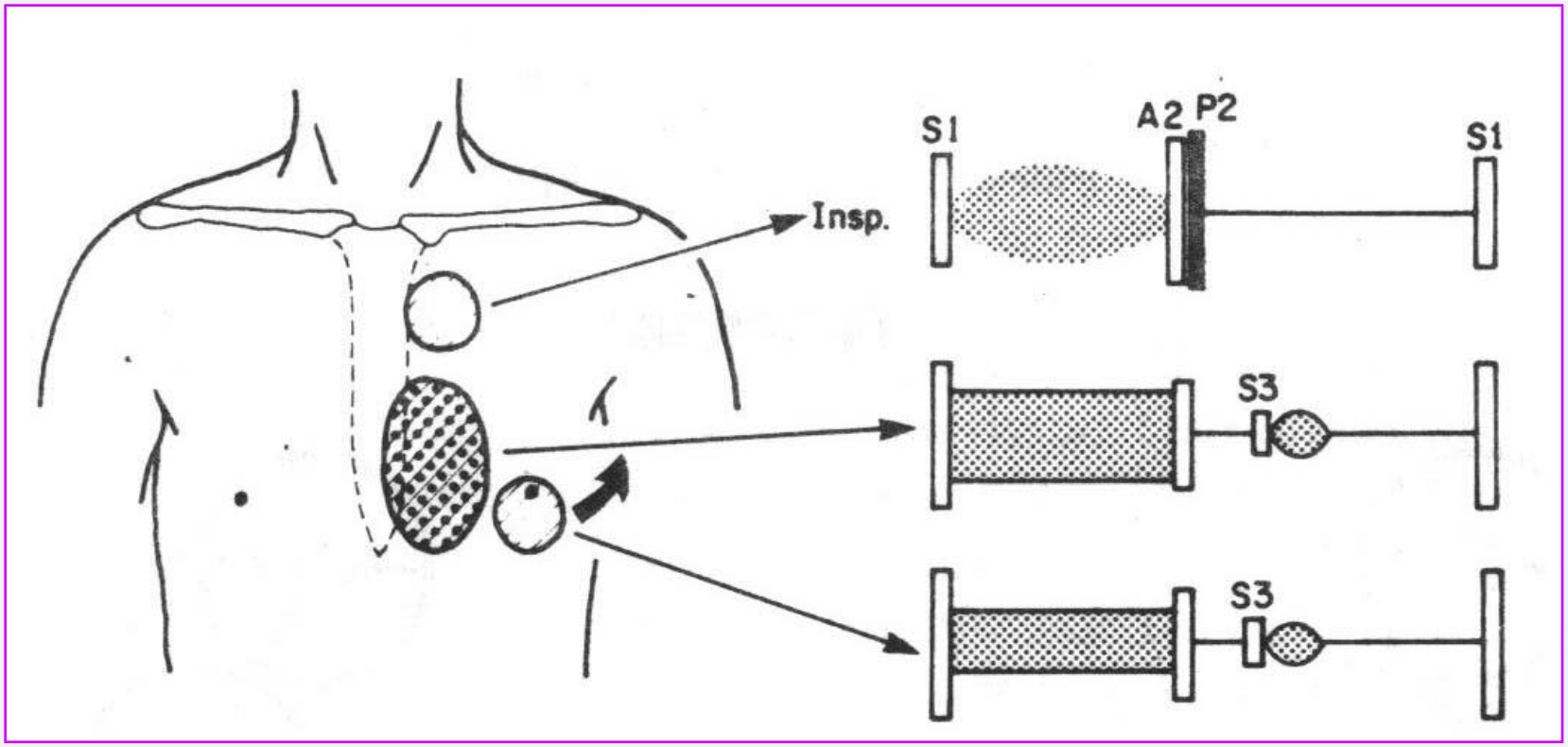
Erken bebeklikte

Kalp yetersizliği

Gelişme geriliği

Sık akciğer enfeks

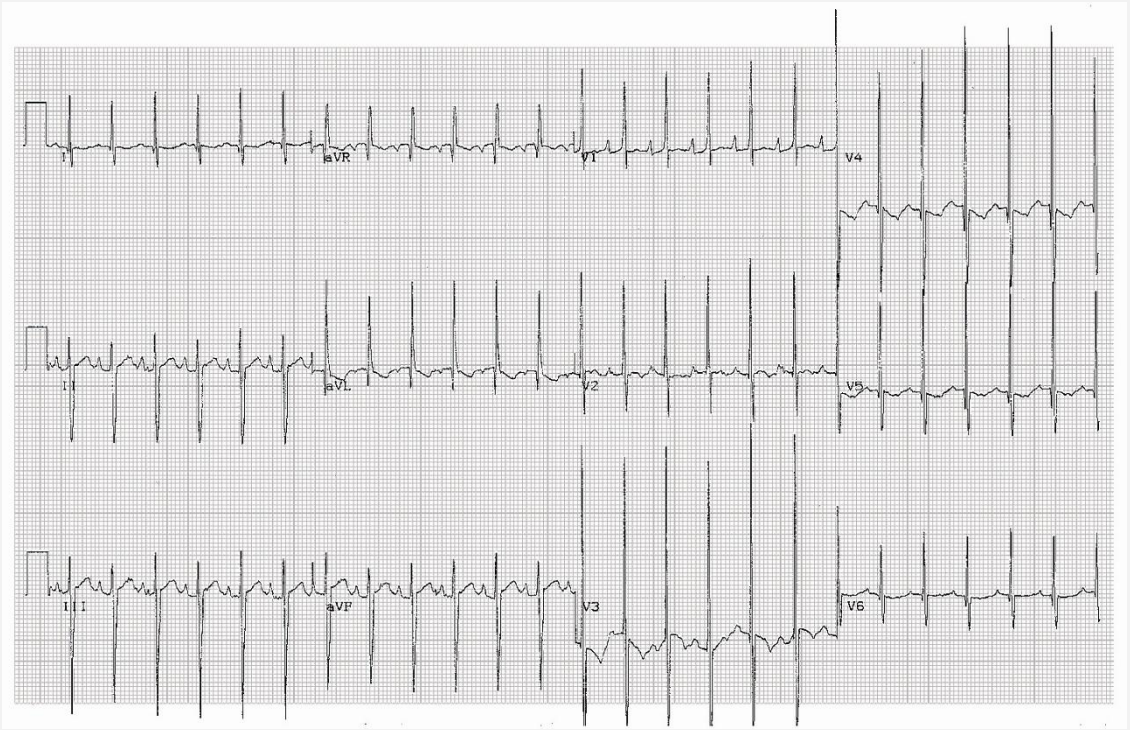
AVSD FİZİK İNCELEME



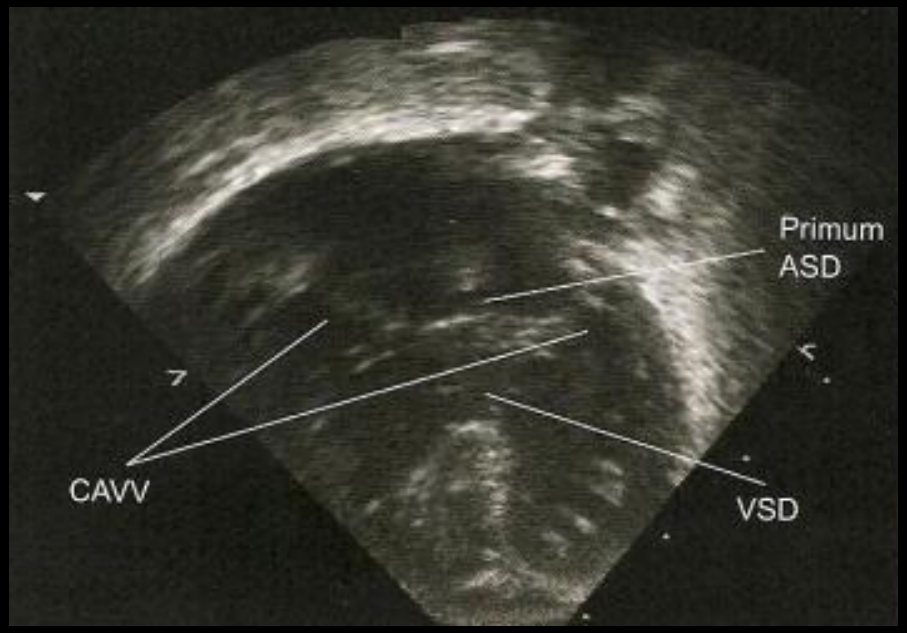
AVSD

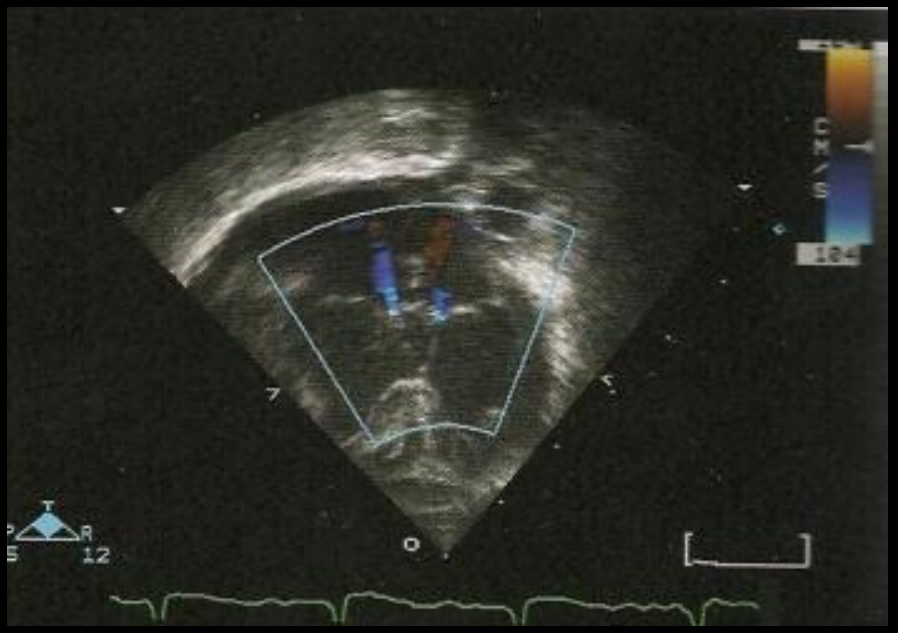
Tele: Kalp büyük
Pulmoner konus belirgin
Akciğer kanlanması artmış

EKG: Sol aks sapması (Sol ant. hemiblok)
Biventriküler hipertrofi, iki atriyumda genişleme
Sağ ventrikül hipertrofisi



EKO: Anatomik tanımlama, PH, kapak yetersizliği



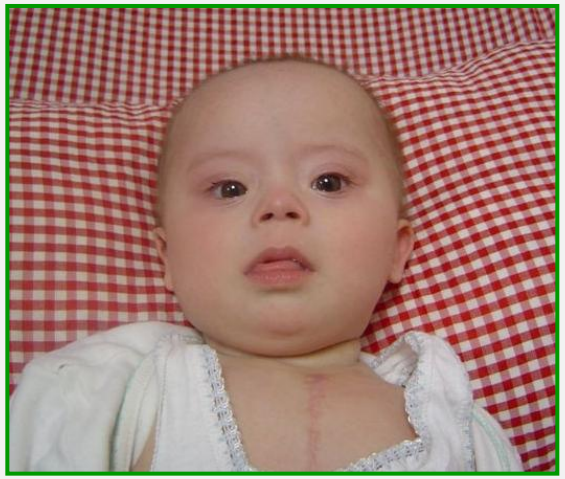


AVSD

Gidiş: Erken kalp yetersizliği

Erken geri dönüşsüz PH

İnfektif endokardit



Tedavi: Kalp yetersizliği tedavisi

< 1 yaş (gnl < 6 ay) CERRAHİ