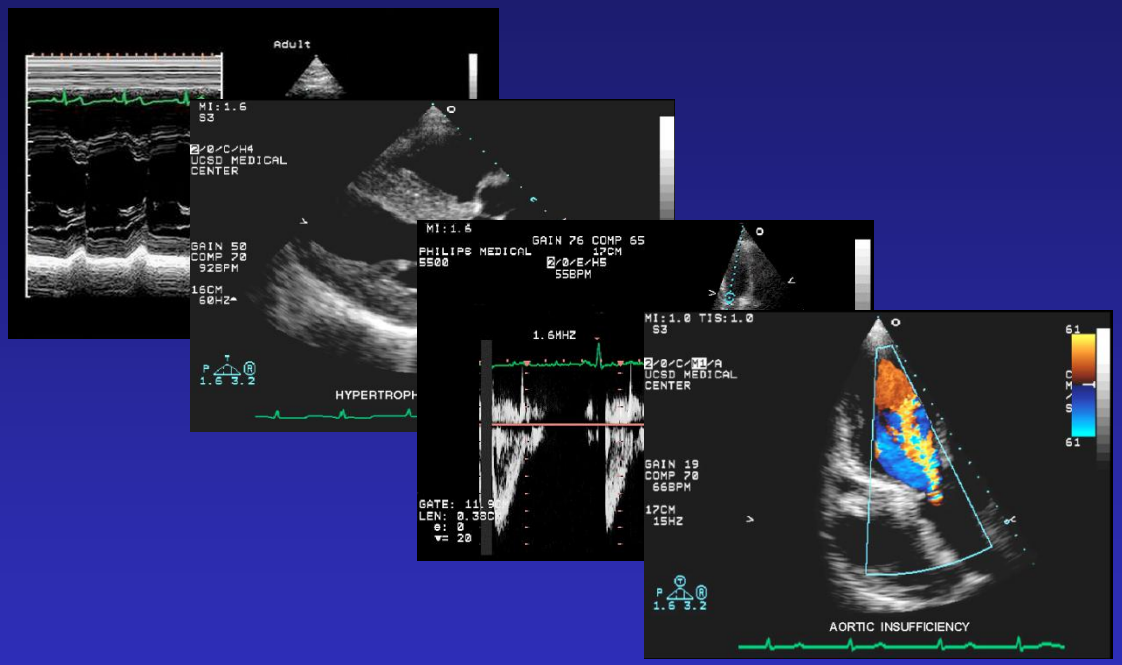
PH pri DPS u pacientov vo vysokom veku – kazuistiky



J. Podracký
S.Juhás

VÝCHODOSLOVENSKÝ ÚSTAV SRDCOVÝCH A CIEVNYCH CHORÔB

DPS u pacientov vo vysokom veku

DPS často nediagnostikovaný do vysokého veku

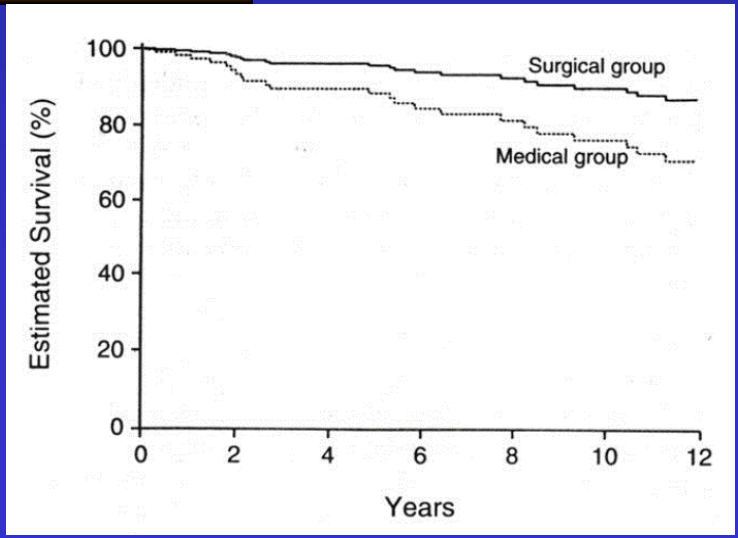
najčastejšie symptómy –

intolerancia námahy /námahová dýchavica ,unavnosť/ palpitácie / poruchy rytmu/

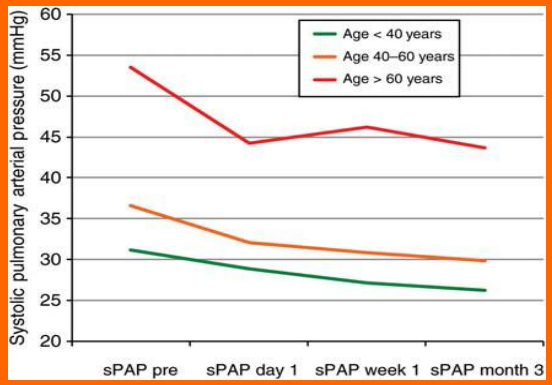
uzáver DPS v dospelosti je kontroverzný -
asymptomatickí pacienti
pacienti so závažnou PH

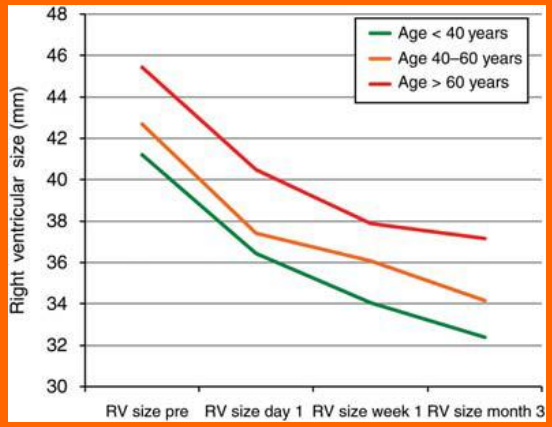
A comparison of surgical and medical therapy for ASD in adults. Konstantinides et al , NEJM, 1995

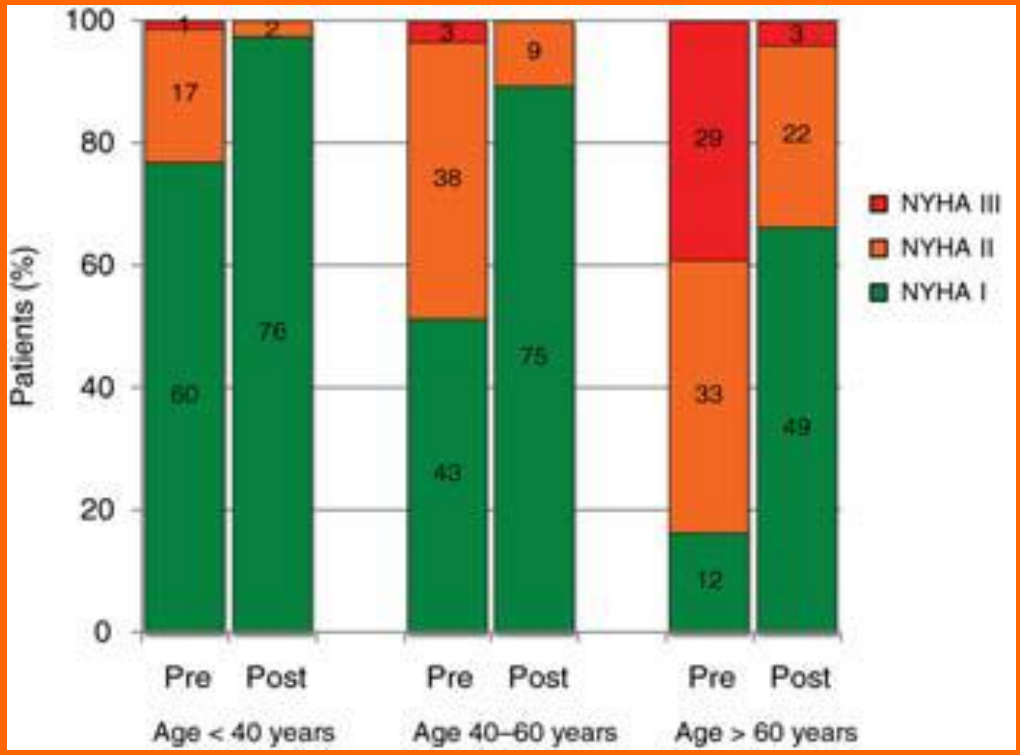
- Retrospective non-randomised
–179 patients over 40 years old
–All had Qp/Qs more than 1.5
–163 (91%) secundum defects.
–Surgery (n= 84) and medical treatment (n=95)
–Mean age at surgery: 56.9 yrs,
–Follow-up evaluation mean: 8.9 years (1-26)



Benefit of atrial septal defect closure in adults: impact of age Humenberger, M et al, EHJ 2011







Clinical classification of pulmonary arterial hypertension associated with congenital heart disease

1. Eisenmenger's syndrome

Includes all large intra- and extra-cardiac defects which begin as systemic-to-pulmonary shunts and progress with time to severe elevation of PVR and to reversal (pulmonary-to-systemic) or bidirectional shunting; cyanosis, secondary erythrocytosis, and multiple organ involvement are usually present.

2. PAH associated with prevalent systemic-to-pulmonary shunts

- Correctable[a]
- Non-correctable

Includes moderate to large defects; PVR is mildly to moderately increased, systemic-to-pulmonary shunting is still prevalent, whereas cyanosis at rest is not a feature.

3. PAH with small/coincidental[b] defects

Marked elevation in PVR in the presence of small cardiac defects (usually ventricular septal defects <1 cm and atrial septal defects <2 cm of effective diameter assessed by echo), which themselves do not account for the development of elevated PVR; the clinical picture is very similar to idiopathic PAH. Closing the defects is contra-indicated.

4. PAH after defect correction

Congenital heart disease is repaired, but PAH either persists immediately after correction or recurs/develops months or years after correction in the absence of significant postoperative haemodynamic lesions.

PAH = pulmonary arterial hypertension; PVR = pulmonary vascular resistance.
[a]With surgery or intravascular percutaneous procedure.
[b]The size applies to adult patients. However, also in adults the simple diameter may be not sufficient for defining the haemodynamic relevance of the defect, and also the pressure gradient, the shunt size and direction, & the pulmonary to systemic flows ratio should be considered
(Web Table II on the web at; www.escardio.org/guidelines).

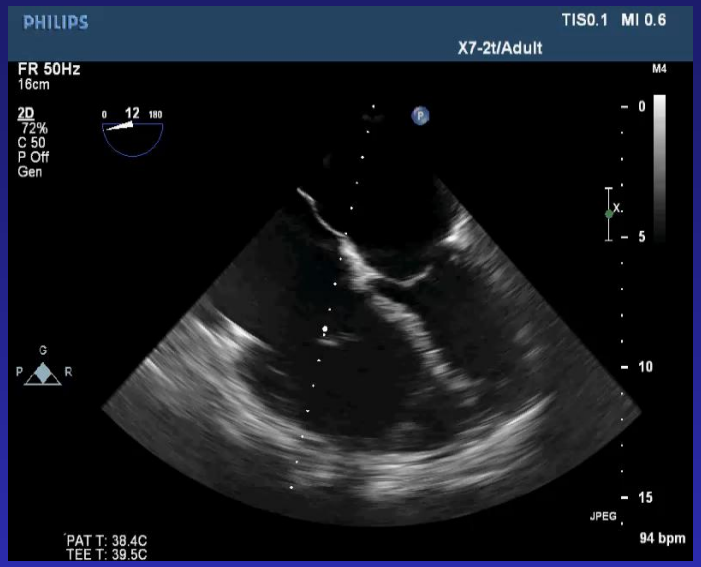
European Heart Journal 2016:37;67–119 -doi:10.1093/eurheartj/ehv317
European Respiratory Journal 2015 46: 903-975;

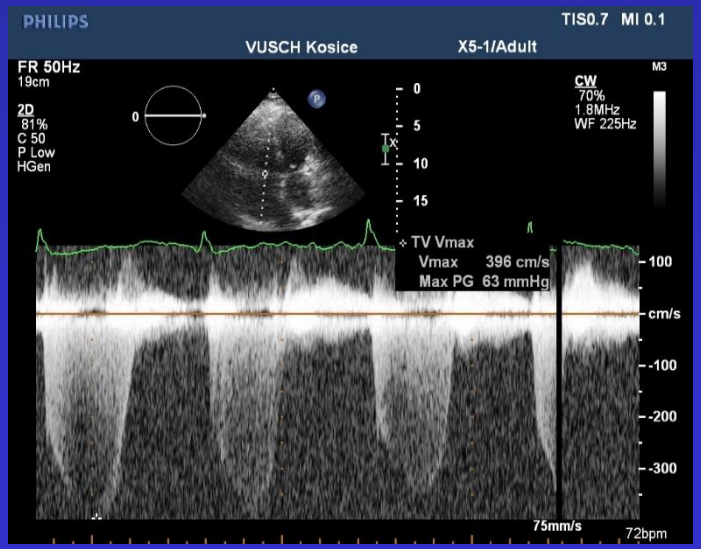
DPS sek. –77r. žena - AA + TTE

2015 – vyšetrovaná pre dyspnoe
/ A – FR v detstve, AH, FP , hypotyreóza/
TTE – dilatácia pravých oddielov, bez PH

1/2017 progresia dýchavice pri námahe
4/2017 HRCT pľúca – iba znaky PH
5/2017 V/P skén – bez poruchy perfuzie

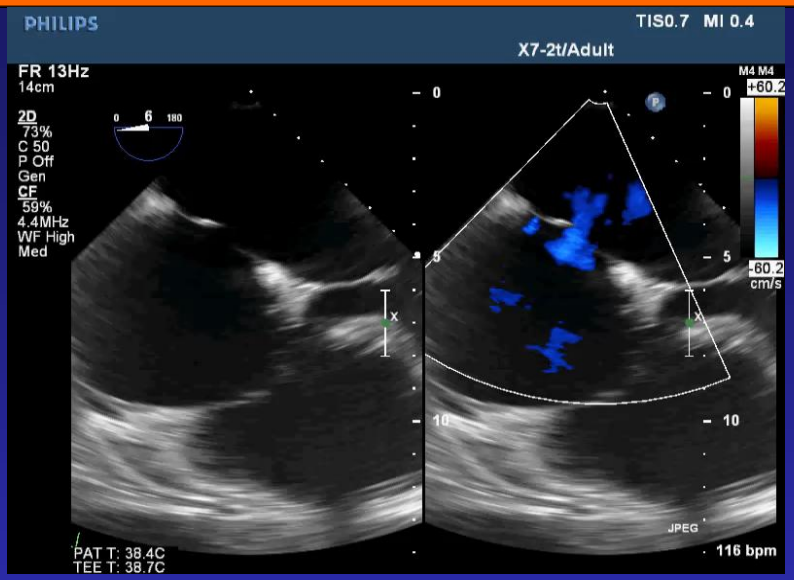
5/2017 TTE- dilat. pravé oddiely,
PK objem. /tlak.preťažená
odhad PASP 60-65 mmHg

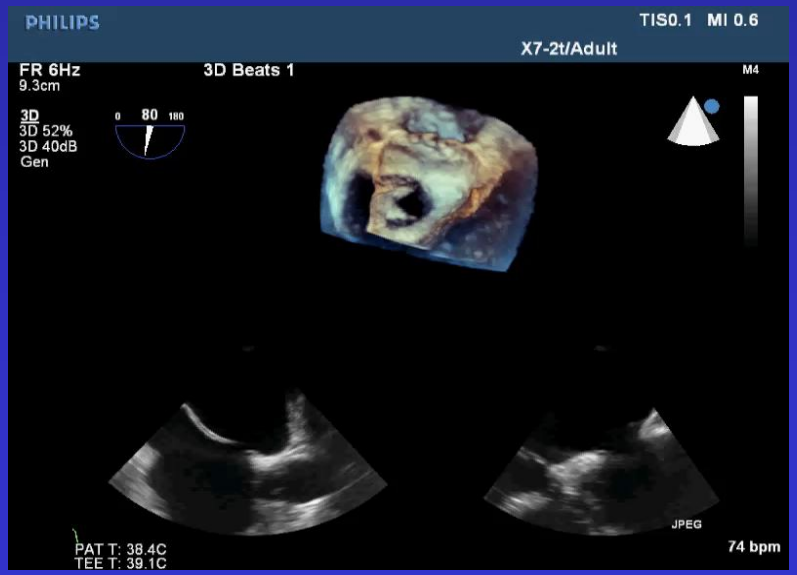


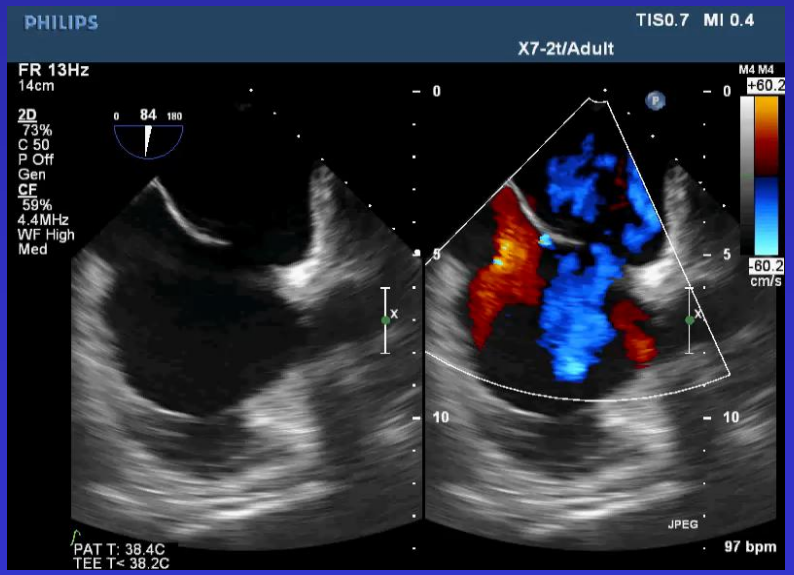


DPS sek. –77r. Žena - TEE

5/2017 TEE-
duplexný DPS centrálne 1,5x1,8cm
menší vzadu 0,4 cm
/ dg. vo veku 76 r. /







DPS sek. –77r. žena - PKS

PKS 05/2017

BSA - 1,8m2

TK 130/70/105 mmHg

PAP 63/30/43 mmHg, RAP 10mmHg

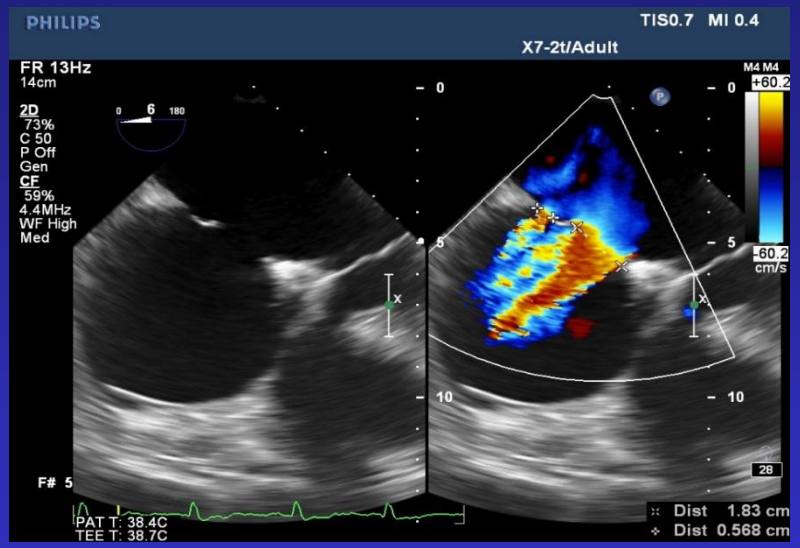
CI 2.7 l/min/m2, PAWP 13mmHg

PVR 4,8 Wj, PVRI 8,6 Wj

SVR 1259, pomer PVR/SVR 0,3

Qp/Qs 3,5

AVT – NO – 43 – 38 mmHg



DPS sek – 77 r. žena – okluder - priebeh

Dg. katetrizácia 06/2017

15 min test uzáveru DPS

PAP 57/20/31 mmHg – pokles

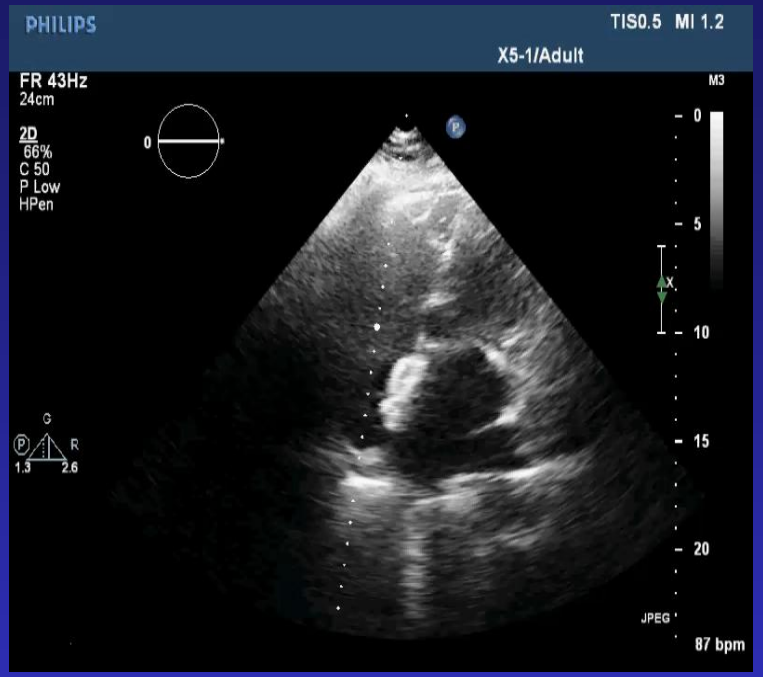
PAWP – bez vzostupu

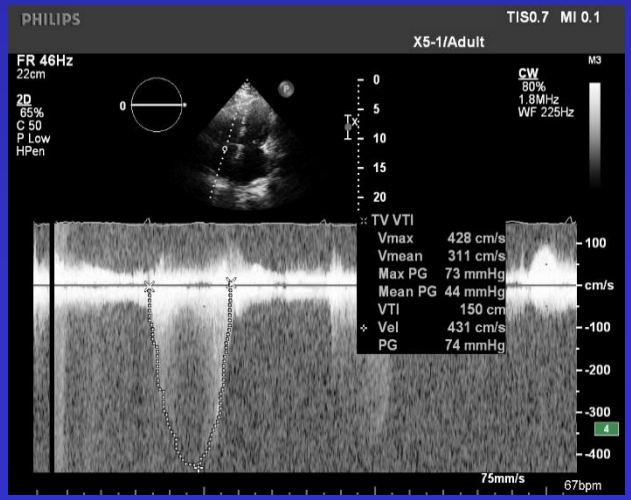
6/2017 Amplatz okluder N22

3/2018 - klinické zhoršenie

– echokg PH idem

7/2018 -ERA - opsumit





Intervencia pri ASD a skratových GUCH

ESC GUCH Guidelines, 2010	PVR < 5 WU with significant shunt regardless of symptoms (class I)	PVR ≥ 5 WU but < 2/3rd SVR, or PAP < 2/3rd systemic blood pressure (baseline or with vasodilator) and net L-to-R shunt (Q_P:Q_S > 1.5; class IIb)	Eisenmenger physiology (class III)
AHA/ACC ACHD Guidelines, 2008	All with RA or RV enlargement, with or without symptoms (class I; no mention of PVR)	Net L-to-R shunting, PAP < 2/3rd systemic, PVR < 2/3rd SVR, or when responsive to either pulmonary vasodilatory therapy or test occlusion of the defect (class IIb)	Severe irreversible PAH and no evidence or a L-to-R shunt (class III)
Updated Clinical PH Classification, 2013	PVR < 2.3WU (< 4WUxm$_2$)	PVR 2.3-4.6WU (4-8 WUxm$_2$)	PVR > 4.6 WU (> 8 WUxm$_2$)

Adapted from Opotowsky AR, *et al. Circulation* 2015; 131:200-210

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Pulmonary arterial hypertension associated with adult congenital heart disease

Recommendations			Class	Level
PVRi (Wu · m²)	PVR (Wu)	Correctable[a]		
<4	<2.3	Yes	IIa	C
>8	>4.6	No	IIa	C
4-8	2.3-4.6	Individual patient evaluation in tertiairy centres	IIa	C

PVR = pulmonary vascular resistance.
PVRi = pulmonary vascular resistance inde.
WU = Wood units.
[a]With surgery or intravascular percutaneous procedure.

ERS EUROPEAN RESPIRATORY SOCIETY

EUROPEAN SOCIETY OF CARDIOLOGY

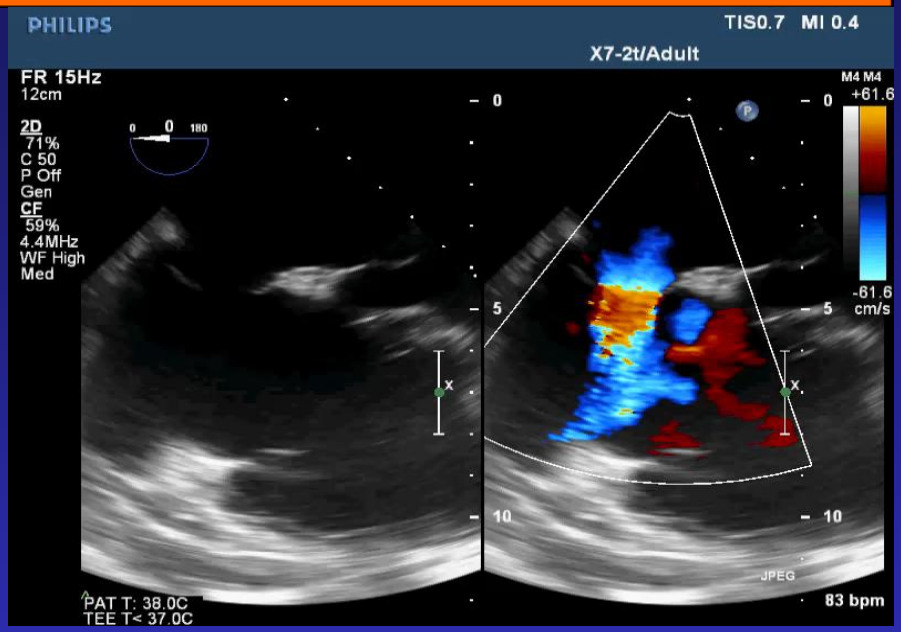
www.escardio.org

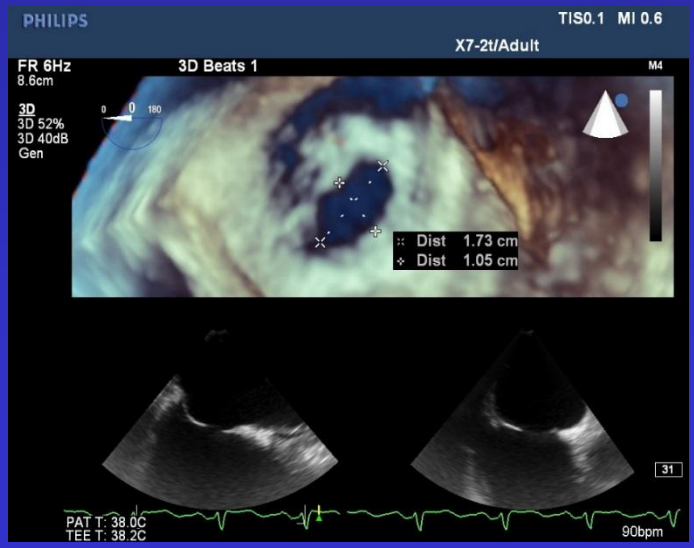
European Heart Journal 2016;37,67–119 -doi:10.1093/eurheartj/ehv317
European Respiratory Journal 2015 46: 903-975;

DPS sek – 72 r. žena – TEE / PKS

2016 – vyšetrovaná pre dyspnoe
/ A – AH, AB , hypotyreóza/

TEE – 05/2017
dilatácia pravých oddielov,
odhad PASP 50 mmHg
DPS sek .1,1x 1,7cm
/ dg. vo veku 71 r/

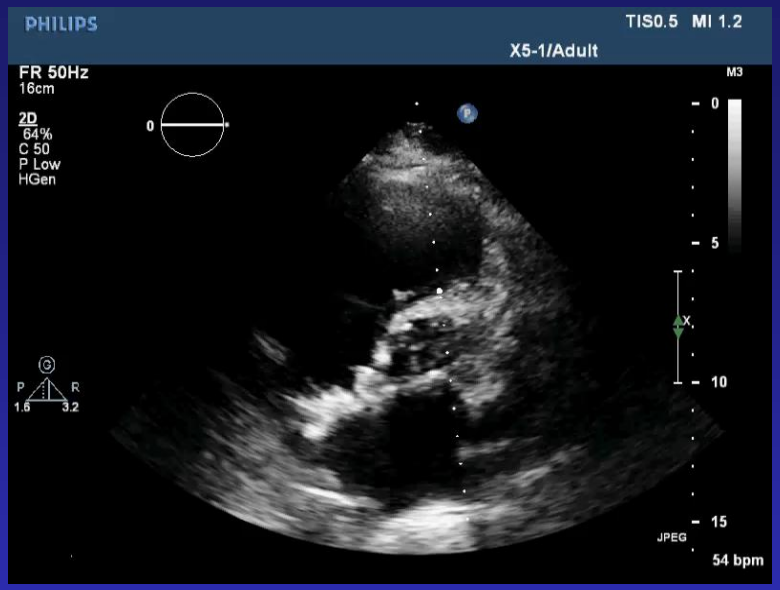


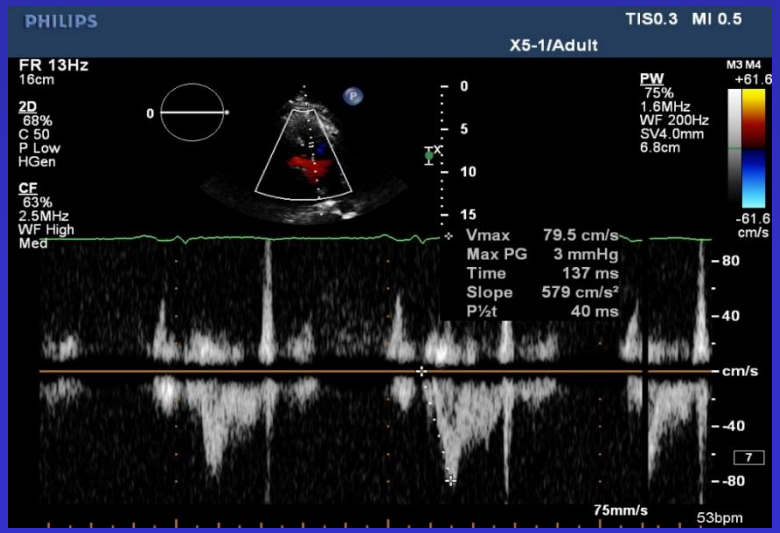


DPS sek – 72 r. žena okluder - priebeh

6/2017 -PKS
PAP 50/15/28 mmHg, PAWP 10
Qp/Qs 3,89

Amplatz okluder N20





Indikácie na uzáver DPS ACC/AHA 2018

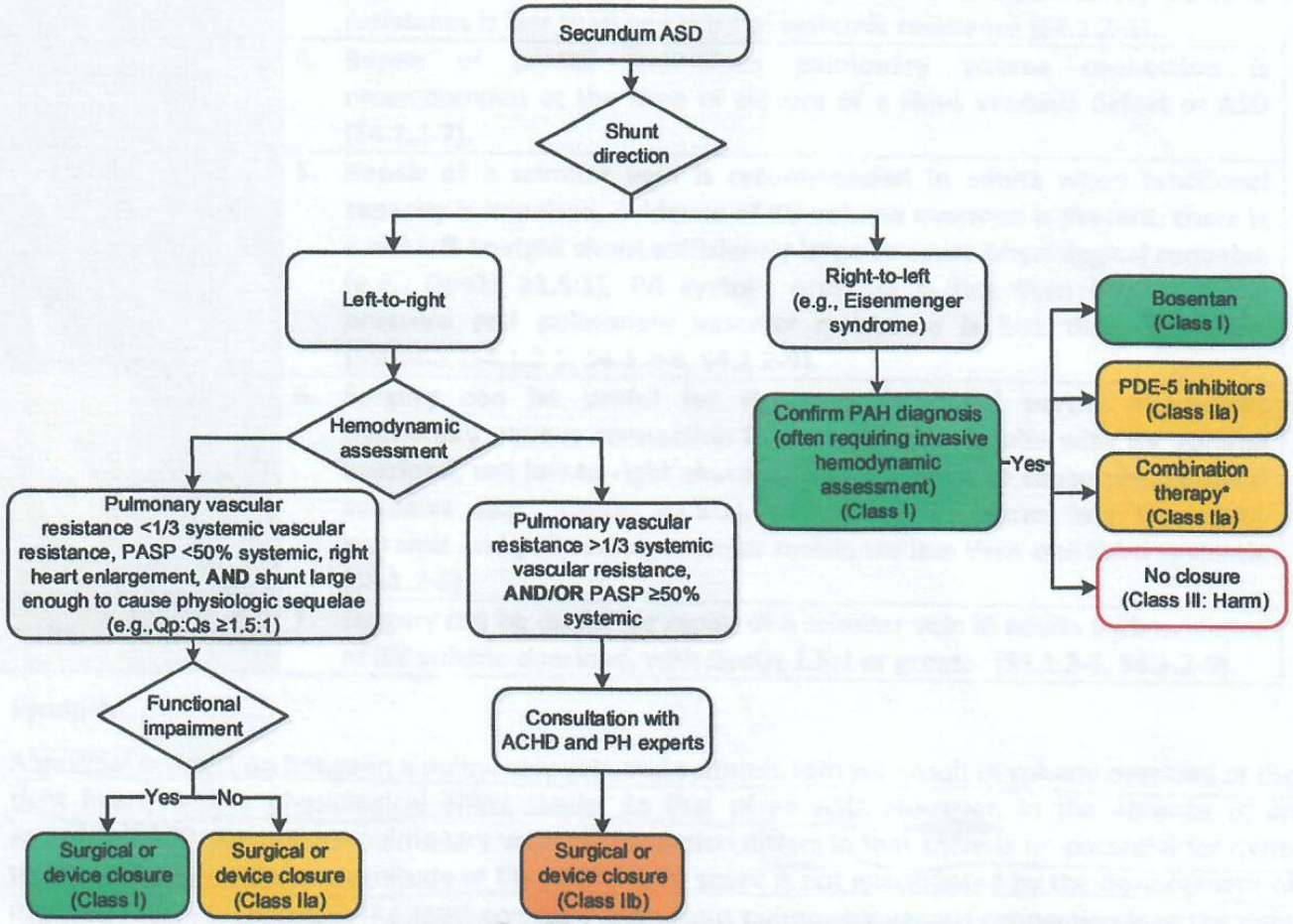
Trieda I
Dospelí s izolovaným DPS sek. so zníženou funkčnou kapacitou, dilatáciou RA a/alevo RV a signifikantným net L-P skratom - Qp:Qs ≥1.5:1, bez cyanózy v pokoji alebo cvičení je odporúčaný transkatétrový alebo KCH uzáver na zlepšenie tolerancie námahy a objem. preťaženia RV, za predpokladu, že systolický PAP je < 50% systémového systolického tlaku a PVR je menej ako 1/3 SVR

Trieda IIa Asymptomatickí dospelí s izolovaným DPS ....

DPS uzáver nemá byť vykonaný u dospelých so systolickým PAP > 2/3 systémového tlaku a PVR > 2/3 SVR a/alebo P-L skratom.

Intervencia pri ASD – ACC/AHA 2018

Figure 1. Secundum ASD



*Combination therapy with bosentan and PDE-5 inhibitor if symptomatic improvement does not occur with either alone.
ACHD indicates adult congenital heart disease; ASD, atrial septal defect; PAH, pulmonary artery hypertension; PASP, pulmonary artery systolic pressure; PDE-5, phosphodiesterase type-5 inhibitors; PH, pulmonary hypertension; and Qp:Qs, pulmonary–systemic blood flow ratio.

DPS vo vysokom veku - závery

uzáver DPS je indikovaný v každom veku
/ zlepšenie symptómov a prognózy, regresia PAP/RV /

preferovaný je katetrizačný uzáver

vo vyššom veku uzáver DPS je treba odporúčať
bez ohľadu na symptómy ihneď po stanovení diagnózy

uzáver nechráni pred predsieň. dysrytmiami
v pokročilom veku problém sú tiež komorbidity

Ďakujem
za pozornosť

Centrum pre dg. a th. PH
VÚSCH a.s. Košice

Indications for intervention in ASD, ESC GUCH, 2010

Patients with significant shunt (signs of RV volume overload) and PVR <5 WU should undergo ASD closure regardless of symptom

Device closure is the method of choice for secundum ASD closure when applicable

All ASDs regardless of size in patients with suspicion of paradox. embolism (exclusion of other causes) should be considered for intervention

Patients with PVR ≥ 5 WU but <2/3 SVR or PAP <2/3 systemic pressure (baseline or when challenged with vasodilators, preferably nitric oxide, or after targeted PAH therapy) and evidence of net L-R shunt (Qp:Qs >1.5) may be considered for intervention

ASD closure must be avoided in patients with Eisenmenger physiology

Pulmonary arterial hypertension associated with congenital heart disease

Recommendations	Class	Level
Bosentan is recommended in WHO-FC III patients with Eisenmenger's syndrome.	I	B
Other ERAs, PDE-5i, and prostanoids should be considered in patients with Eisenmenger's syndrome.	IIa	C
In the absence of significant haemoptysis, oral anticoagulant treatment may be considered in patients with PA thrombosis or signs of heart failure.	IIb	C
The use of supplemental O_2 therapy should be considered in cases in which it produces a consistent increase in arterial oxygen saturation and reduces symptoms.	IIa	C
If symptoms of hyperviscosity are present, phlebotomy with isovolumic replacement should be considered, usually when the haematocrit is >65%.	IIa	C
The use of supplemental iron treatment may be considered in patients with low ferritin plasma levels.	IIb	C
Combination drug therapy may be considered in patients with Eisenmenger's syndrome.	IIb	C
The use of CCBs is not recommended in patients with Eisenmenger's syndrome.	III	

European Heart Journal 2016:37;67–119 -doi:10.1093/eurheartj/ehv317
European Respiratory Journal 2015 46: 903-975;

ASD : atrial arrhythmias

- Atrial arrhythmias increased with age
- preoperative atrial flutter or paroxysmal supraventricular tachycardia can evolve into sustained postoperative AF
- surgical correction of ASD in adults did not decrease the occurrence of AF.

Left ventricular dysfunction

- elderly and hypertensive
- hypertrophied, less compliant LV.
- Restrictive left ventricular dysfunction in elderly may be masked by the presence of an ASD.
- Deterioration of left ventricular diastolic function can occur with acute hemodynamic change following closure of ASD, leading to acute lung edema.

Pulmonary hypertension

- PHT increases progressively with advancing age
- Rate of progression is variable
- Multifactorial cause
- Uncommon , 5-10% pulmonary vascular disease