Communication InterAuriculaire (CIA)

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Cardiologie pédiatrique

Hôpital Necker Enfants malades

Centre de Référence des Malformations Cardiaques Congénitales Complexes M3C

Epidémiologie

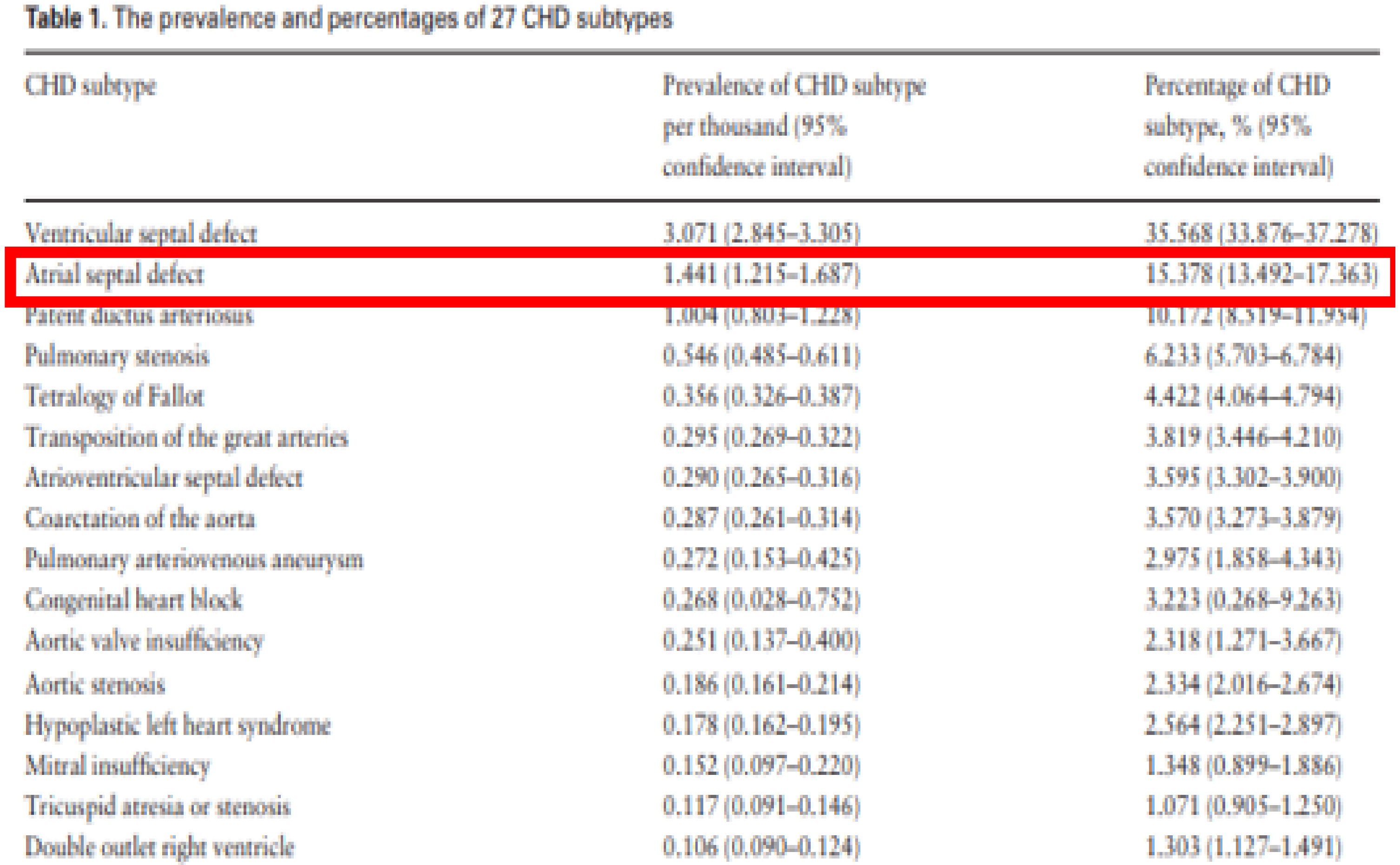
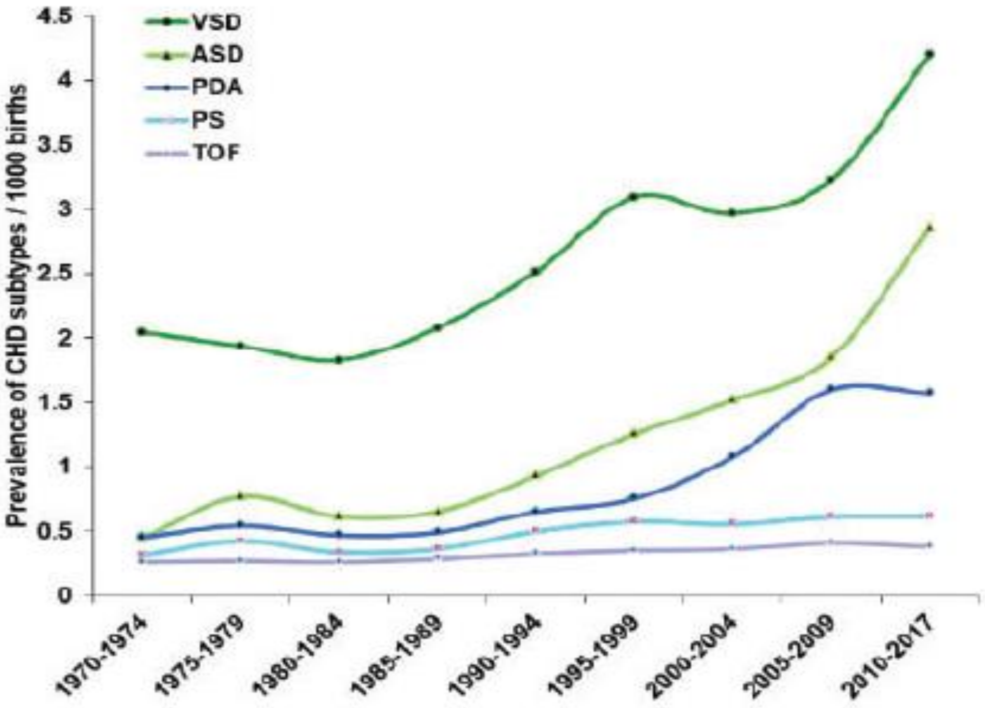


Table 1. The prevalence and percentages of 27 CHD subtypes

CHD subtype	Prevalence of CHD subtype per thousand (95% confidence interval)	Percentage of CHD subtype, % (95% confidence interval)
Ventricular septal defect	3.071 (2.845–3.305)	35.568 (33.876–37.278)
Atrial septal defect	1.441 (1.215–1.687)	15.378 (13.492–17.363)
Patent ductus arteriosus	1.004 (0.803–1.228)	10.172 (8.519–11.954)
Pulmonary stenosis	0.546 (0.485–0.611)	6.233 (5.703–6.784)
Tetralogy of Fallot	0.356 (0.326–0.387)	4.422 (4.064–4.794)
Transposition of the great arteries	0.295 (0.269–0.322)	3.819 (3.446–4.210)
Atrioventricular septal defect	0.290 (0.265–0.316)	3.595 (3.302–3.900)
Coarctation of the aorta	0.287 (0.261–0.314)	3.570 (3.273–3.879)
Pulmonary arteriovenous aneurysm	0.272 (0.153–0.425)	2.975 (1.858–4.343)
Congenital heart block	0.268 (0.028–0.752)	3.223 (0.268–9.263)
Aortic valve insufficiency	0.251 (0.137–0.400)	2.318 (1.271–3.667)
Aortic stenosis	0.186 (0.161–0.214)	2.334 (2.016–2.674)
Hypoplastic left heart syndrome	0.178 (0.162–0.195)	2.564 (2.251–2.897)
Mitral insufficiency	0.152 (0.097–0.220)	1.348 (0.899–1.886)
Tricuspid atresia or stenosis	0.117 (0.091–0.146)	1.071 (0.905–1.250)
Double outlet right ventricle	0.106 (0.090–0.124)	1.303 (1.127–1.491)



Yingjuan Liu, Sen Chen, Liesl Zühlke, Graeme C Black, Mun-kit Choy, Ningxiu Li, Bernard D Keavney, Global birth prevalence of congenital heart defects 1970–2017: updated systematic review and meta-analysis of 260 studies, *International Journal of Epidemiology*, Volume 48, Issue 2, April 2019, Pages 455–463, https://doi.org/10.1093/ije/dyz009

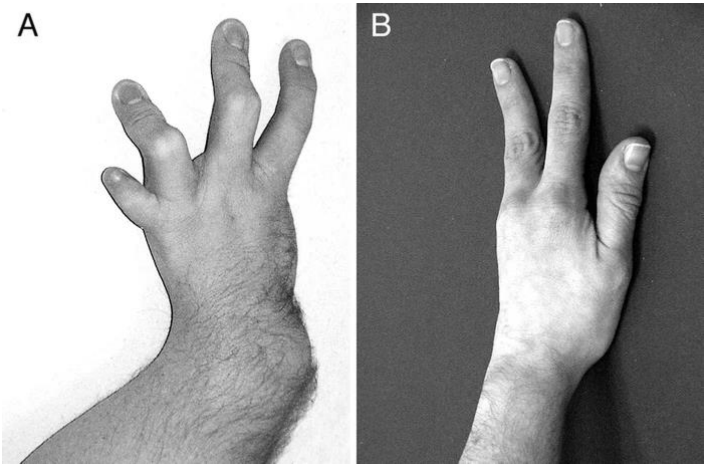
Etiologies des CIA

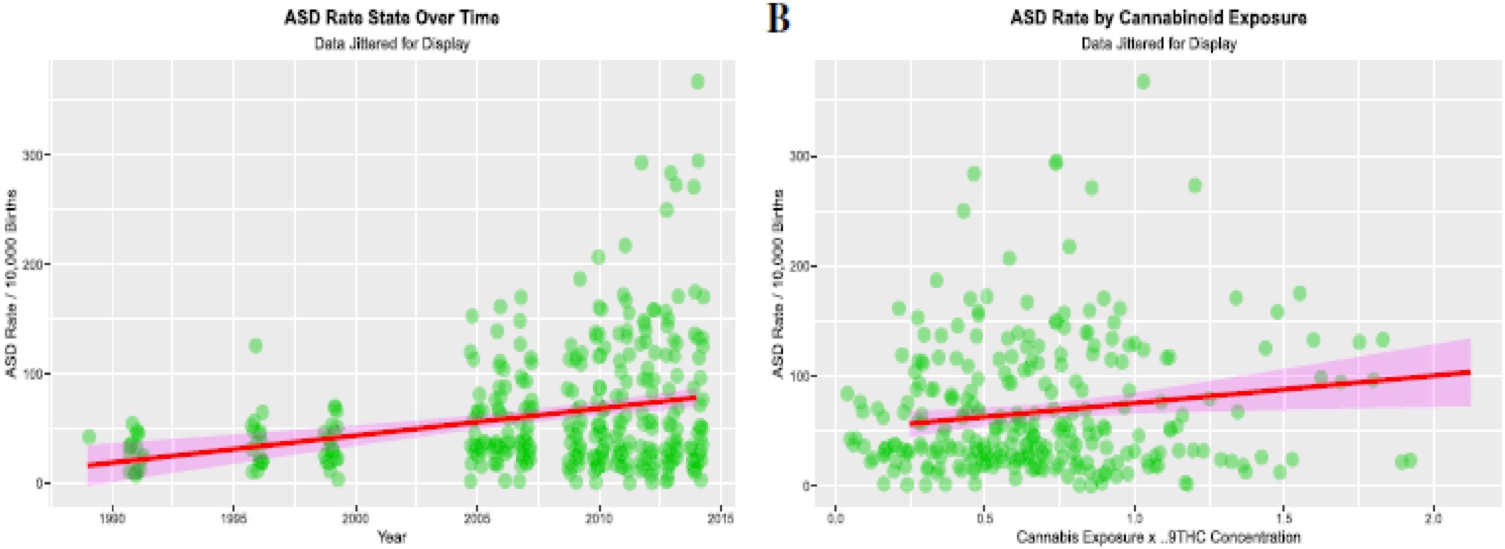
Le plus souvent sporadiques/ idiopathique

Parfois formes familiales associées :

- BAV du 1er degré : gène Nkx2.5
- Syndrome de Holt-Oram
- Downs syndrome
- GATA4, Tbx5, TGFβ

❖ Environnement : Cannabis

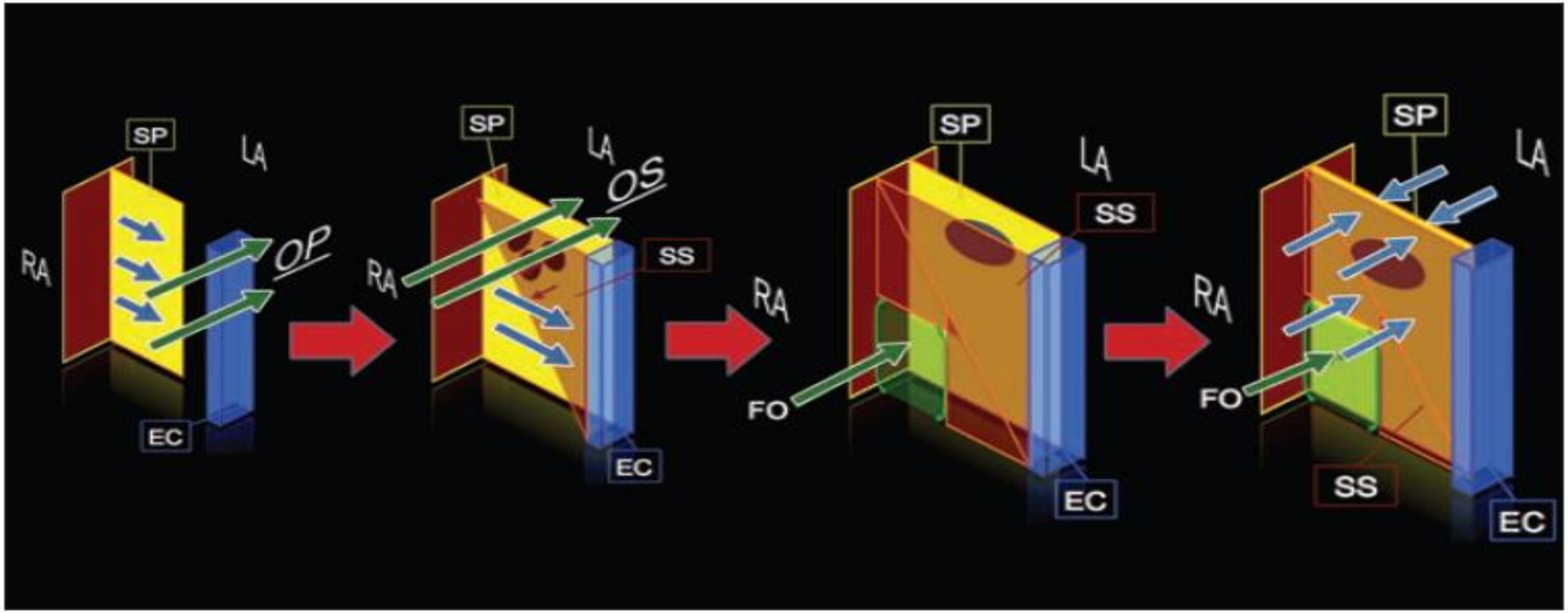




Reece, A.S., Hulse, G.K. Contemporary epidemiology of rising atrial septal defect trends across USA 1991–2016: a combined ecological geospatiotemporal and causal inferential study. *BMC Pediatr* **20,** 539 (2020). https://doi.org/10.1186/s12887-020-02431-z

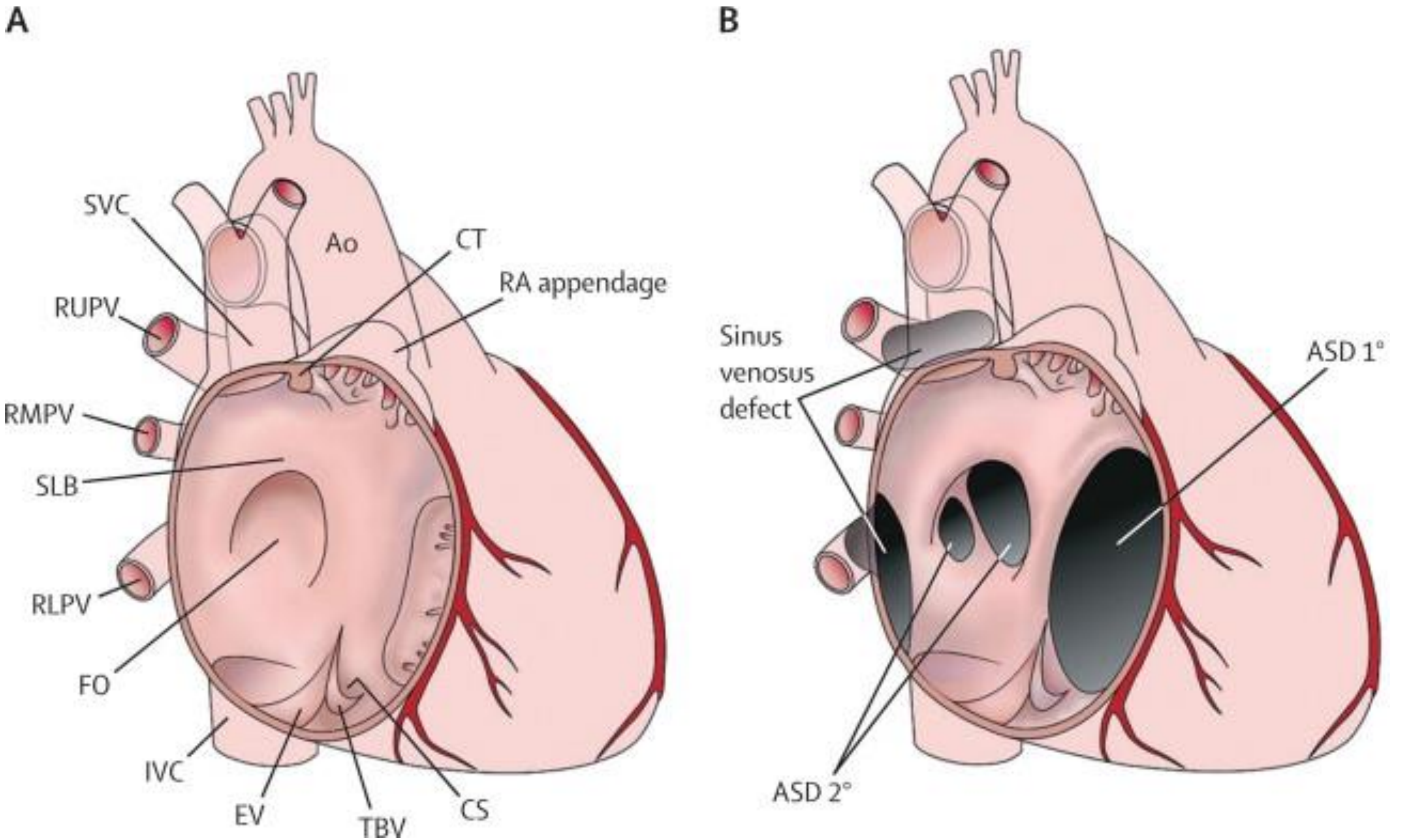
Posch, M.G., Perrot, A., Berger, F. *et al.* Molecular genetics of congenital atrial septal defects. *Clin Res Cardiol* **99,** 137–147 (2010). https://doi.org/10.1007/s00392-009-0095-0

Embryologie du SIA


SP
LA
RA
OP
EC
SP
LA
OS
SS
RA
EC
SP
LA
SS
RA
FO
EC
SP
LA
RA
FO
SS
EC

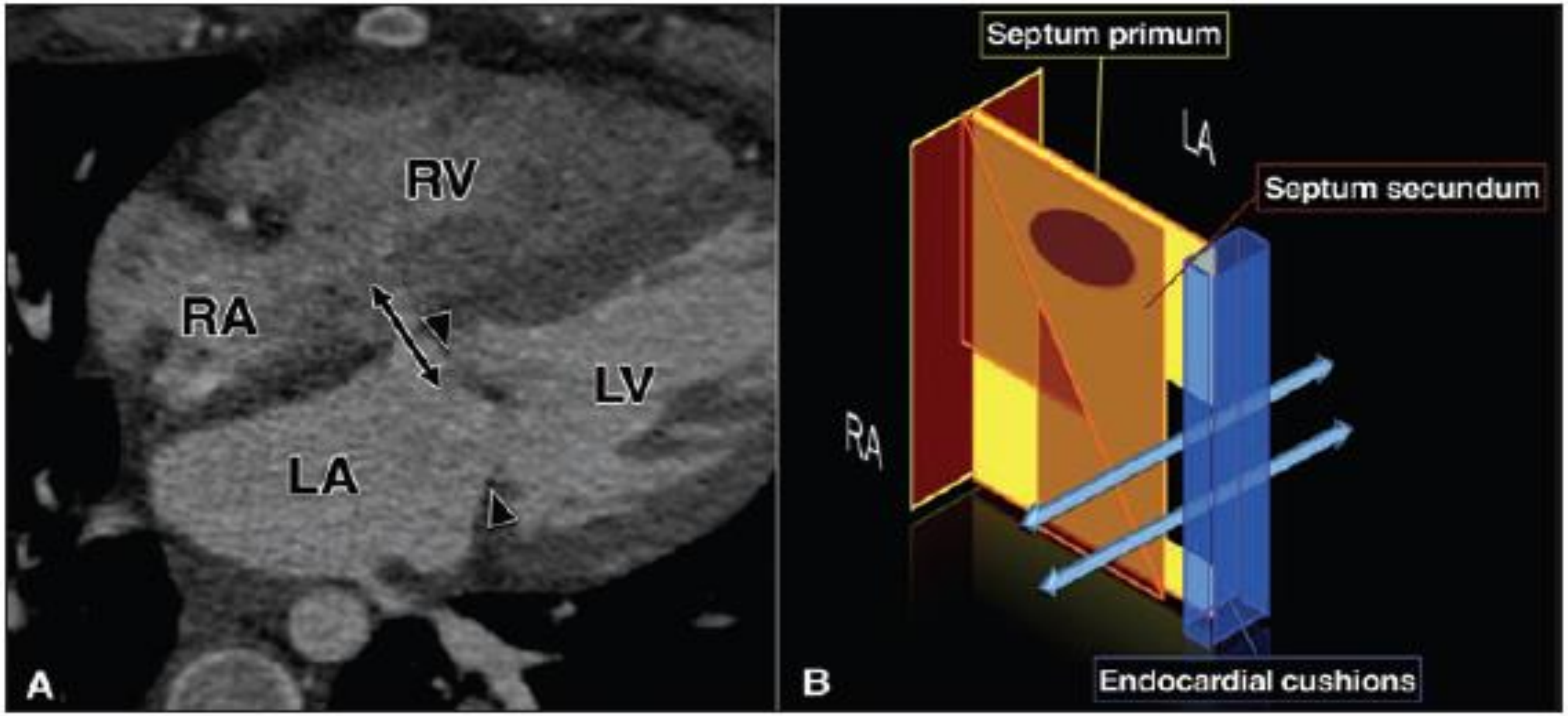
Rojas *et al*. AJR 2010

Anatomie des CIA


A
B
SVC
Ao
CT
RA appendage
RUPV
RMPV
SLB
RLPV
FO
IVC
EV
TBV
CS
Sinus venosus defect
ASD 1°
ASD 2°

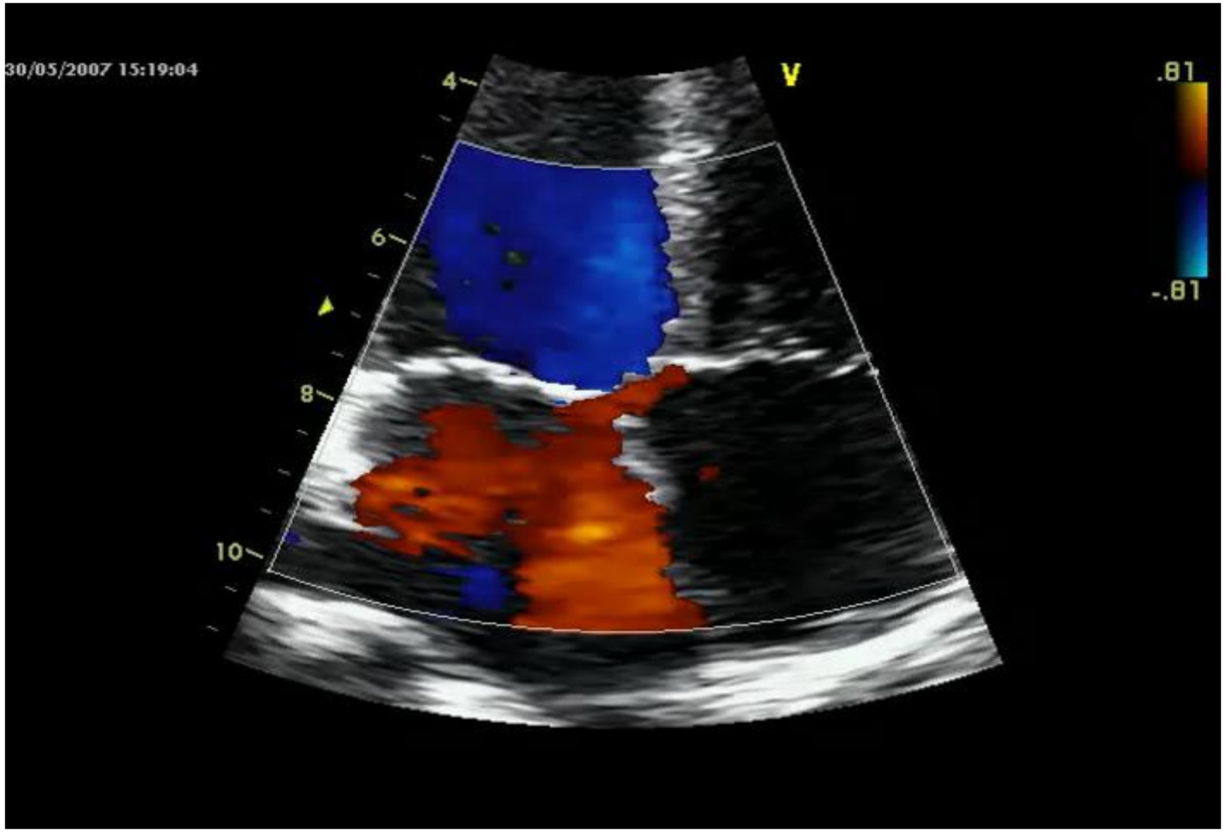
CIA Ostium Primum

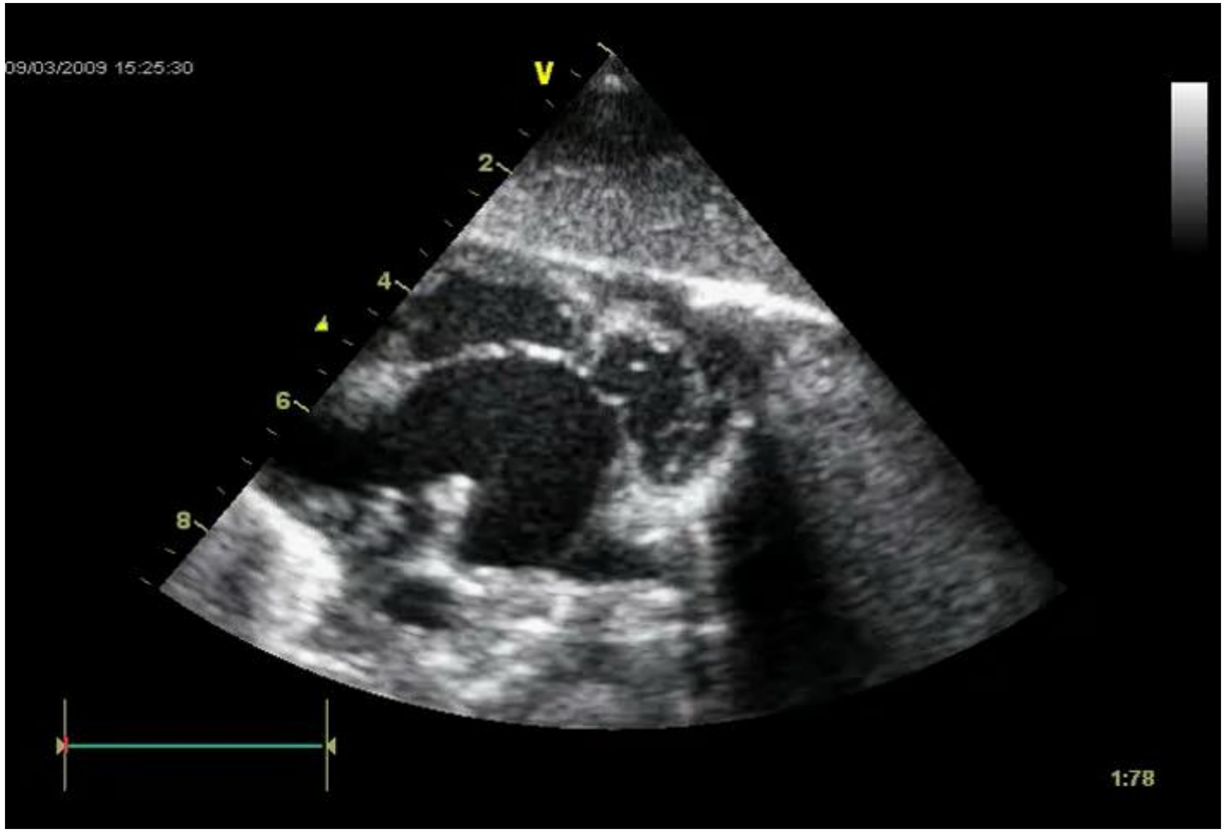
- 2–3% of the ASDs
- associated with Down syndrome


RV
RA
LV
LA
A
Septum primum
LA
Septum secundum
RA
B
Endocardial cushions

Rojas *et al*. AJR 2010

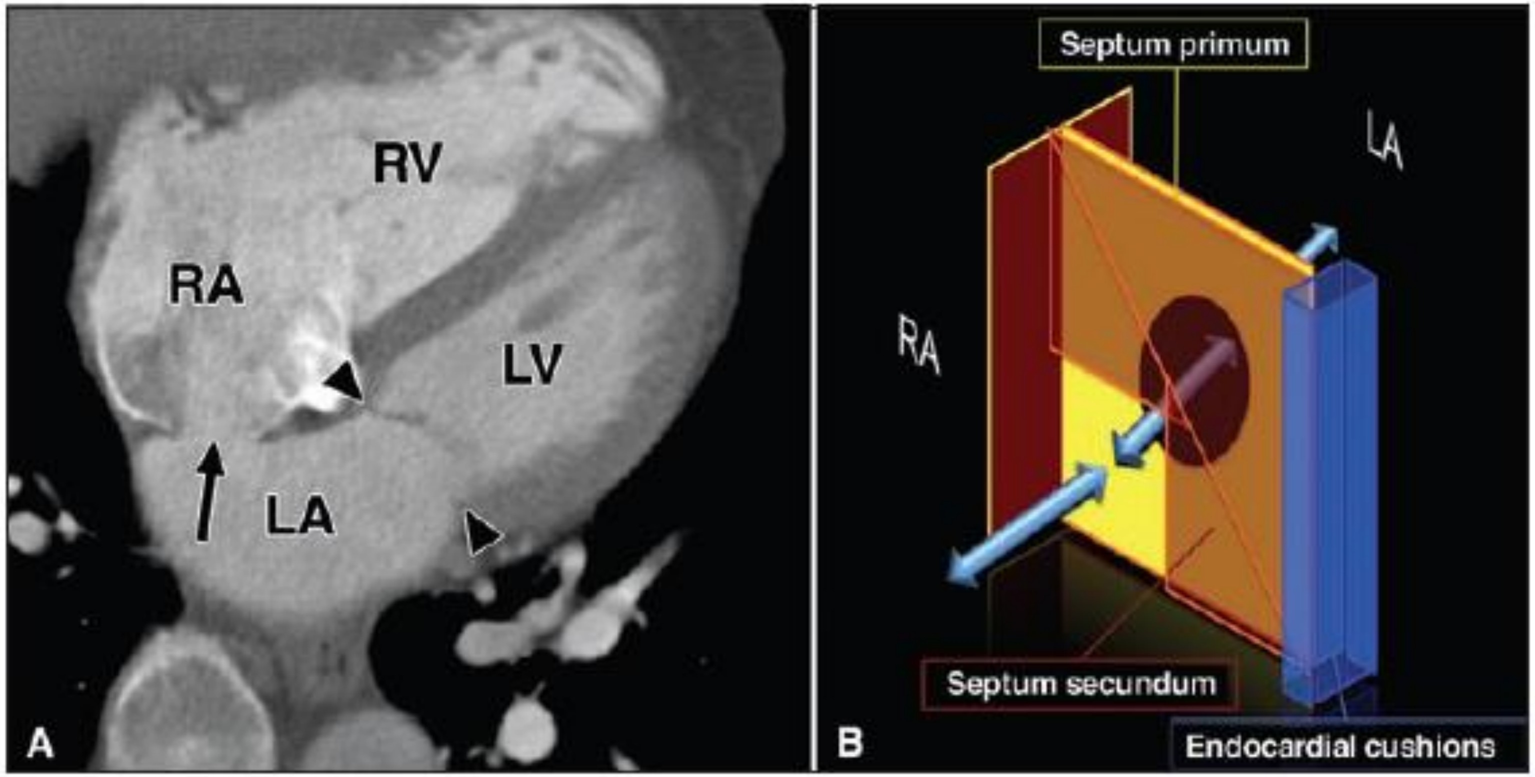
CIA Ostium Primum


30/05/2007 15:19:04
V
.81
-.81
4
6
8
10


09/03/2009 15:25:30
V
2
4
6
8
1:78

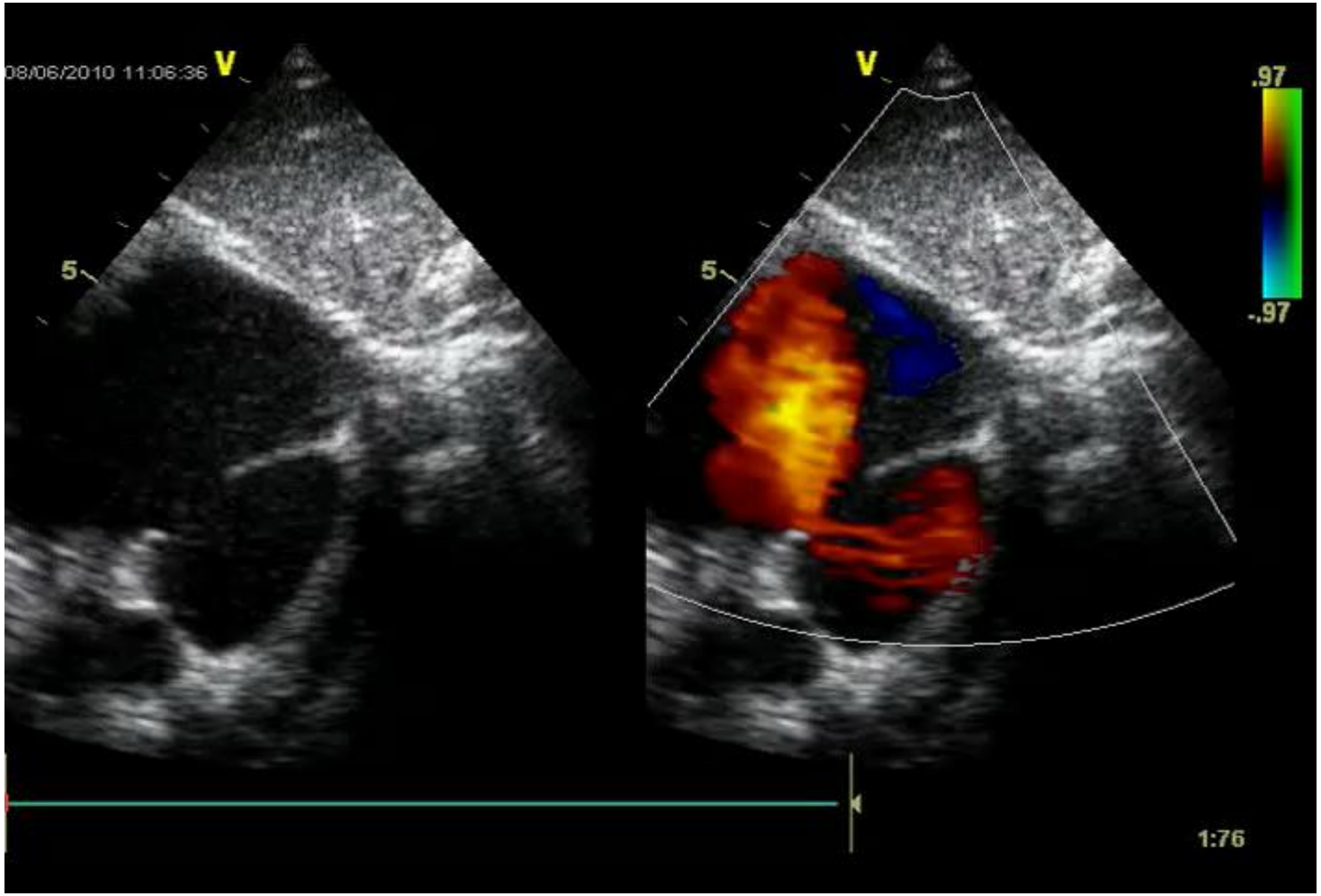
CIA Ostium Secundum

80-90 % des CIAs


RV
RA
LV
LA
A
Septum primum
LA
RA
Septum secundum
B
Endocardial cushions

Rojas *et al*. AJR 2010

CIA Ostium Secundum

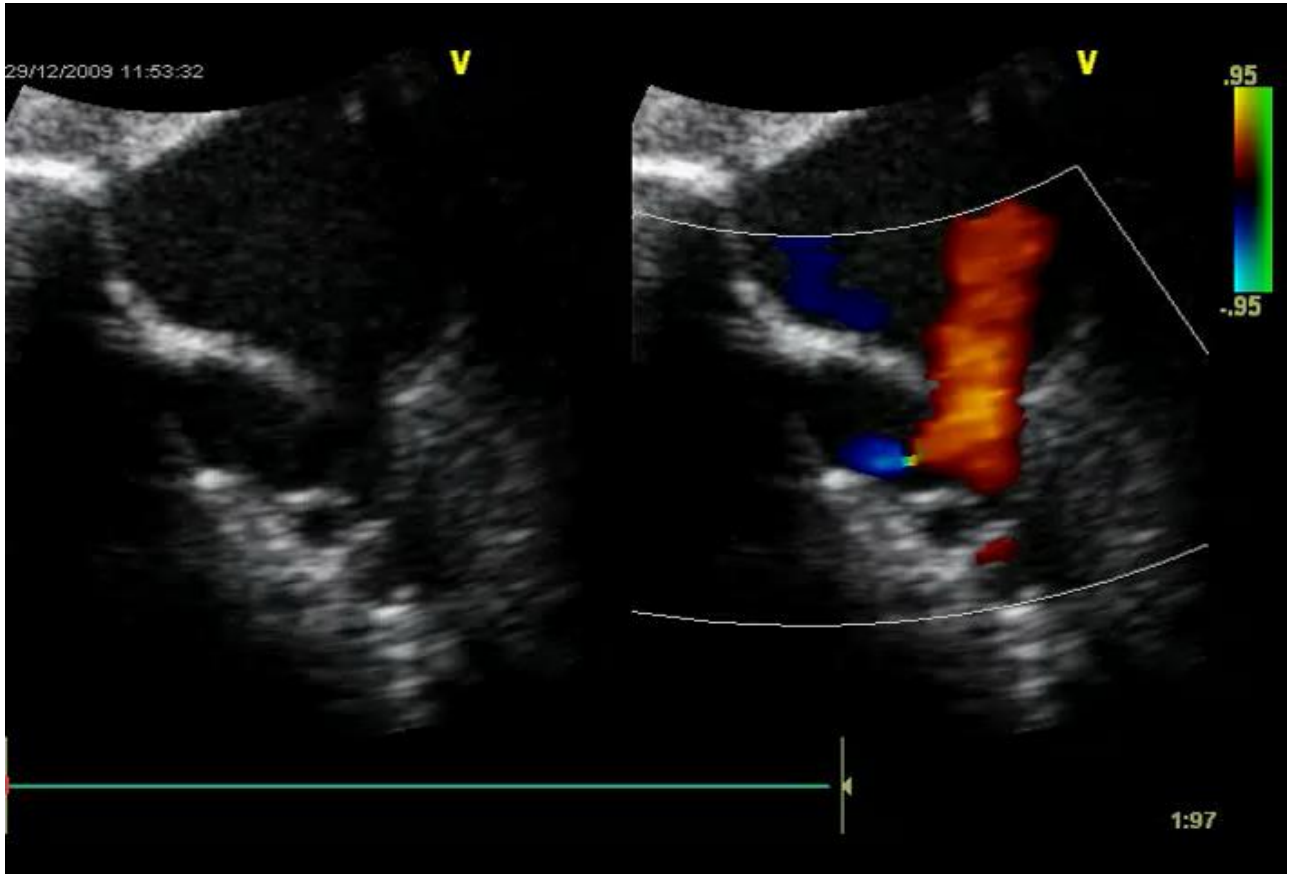

08/06/2010 11:06:36
V
5
V
5
.97
-.97
1:76

CIA Sinus venosus

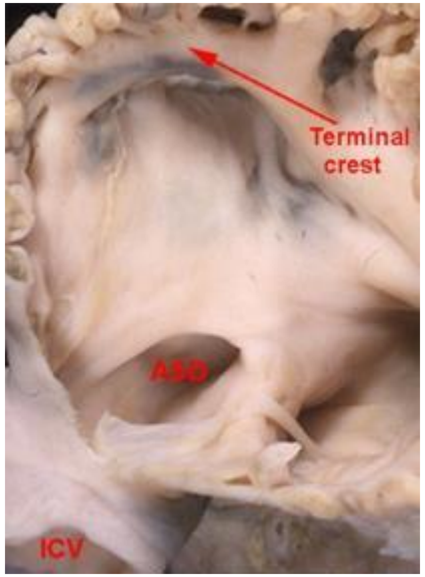
2–10%
Embryologie controversée

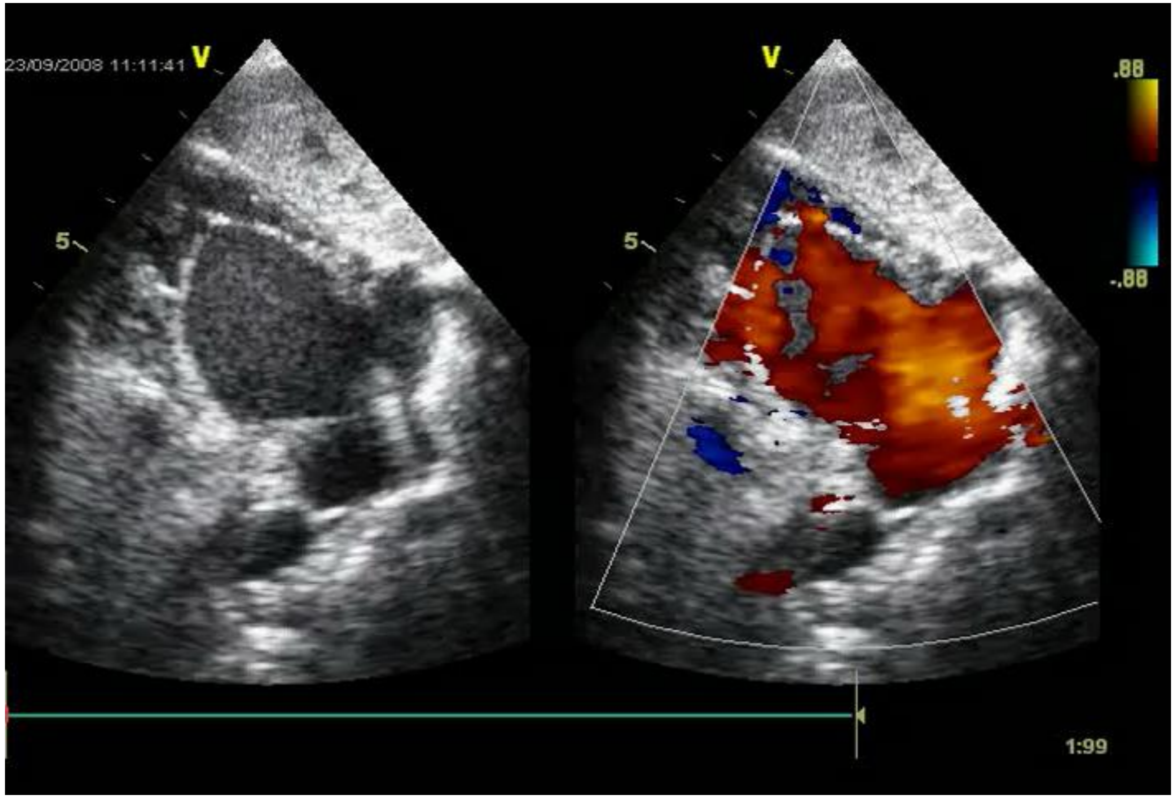
La CIA du SV supérieur est un défaut de communication entre l’ OD et la jonction VCS - OG.
Souvent associé à un RVPAp du LSD dans la VCS.

La CIA du SV inférieur est un défaut de communication entre l’ OD et la jonction VCI - OG.
Peut être associé à un RVPAp du LID

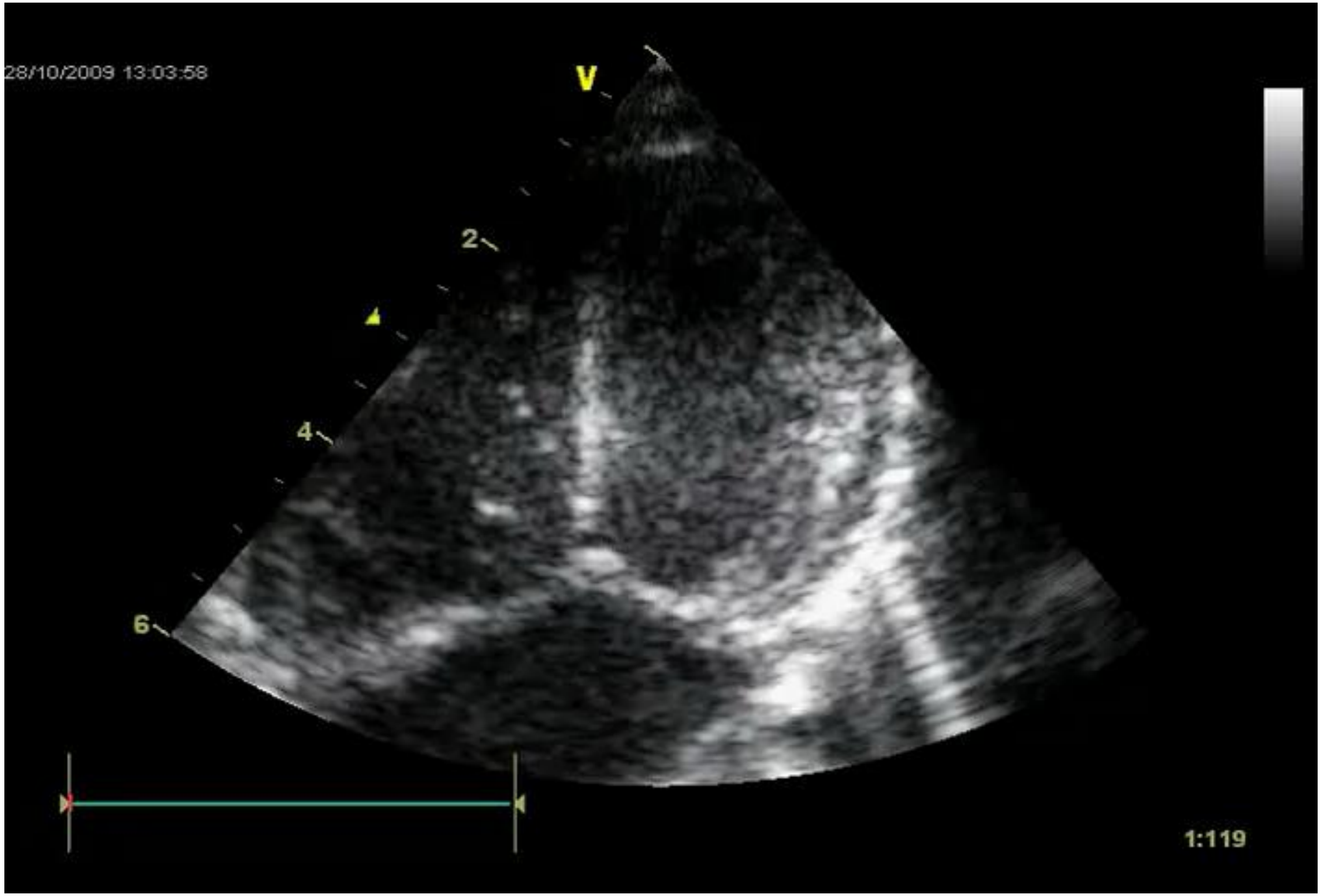


CIA low septal defect ou CIA basse


Terminal crest
ASD
ICV

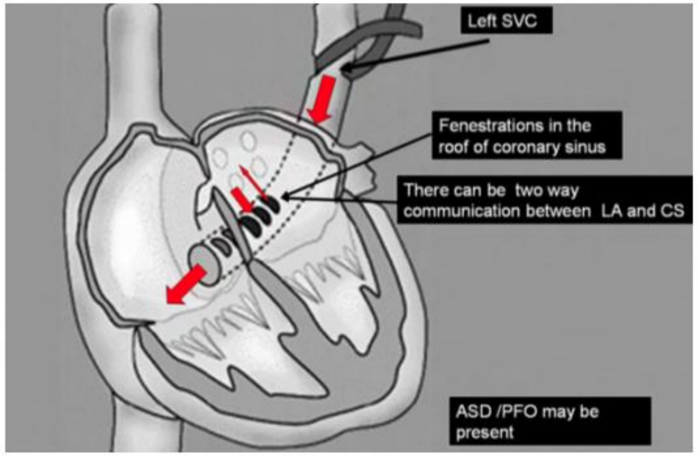

23/09/2008 11:11:41
V
5
V
5
.88
-.88
1:99

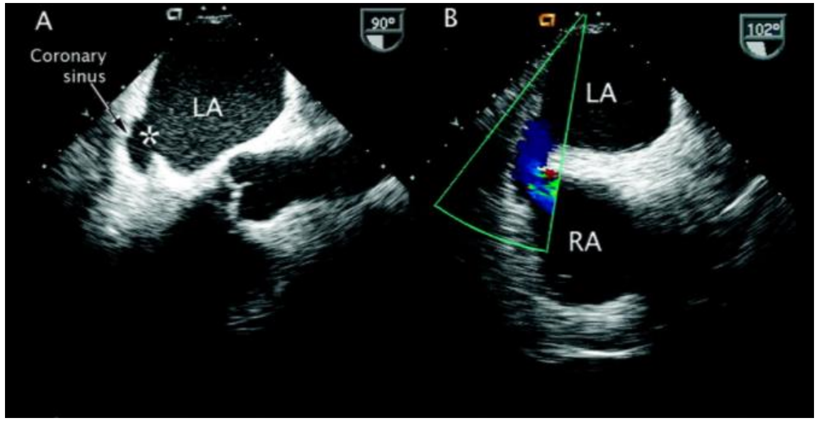
Oreillette unique

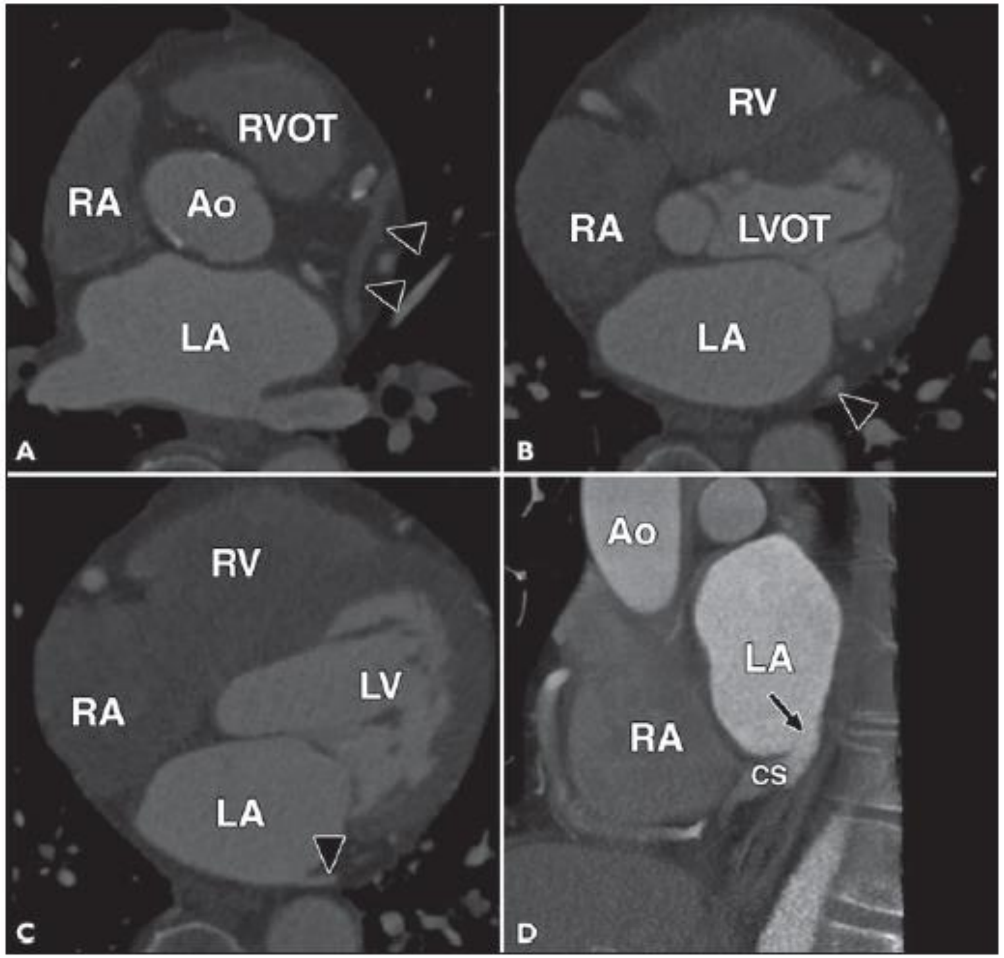

28/10/2009 13:03:58
V
2
4
6
1:119

CIA du sinus coronaire

< 1%
Absence de septation entre l’OG inférieure et le toit du SC
Fréquemment associé à une VCSG


Left SVC
Fenestrations in the roof of coronary sinus
There can be two way communication between LA and CS
ASD /PFO may be present


A
Coronary sinus
*
LA
90°
B
LA
RA
102°


RVOT
RA
Ao
LA
A
RV
RA
LVOT
LA
B
RV
RA
LV
LA
C
Ao
LA
RA
CS
D

Physiologie du shunt à l’étage atrial

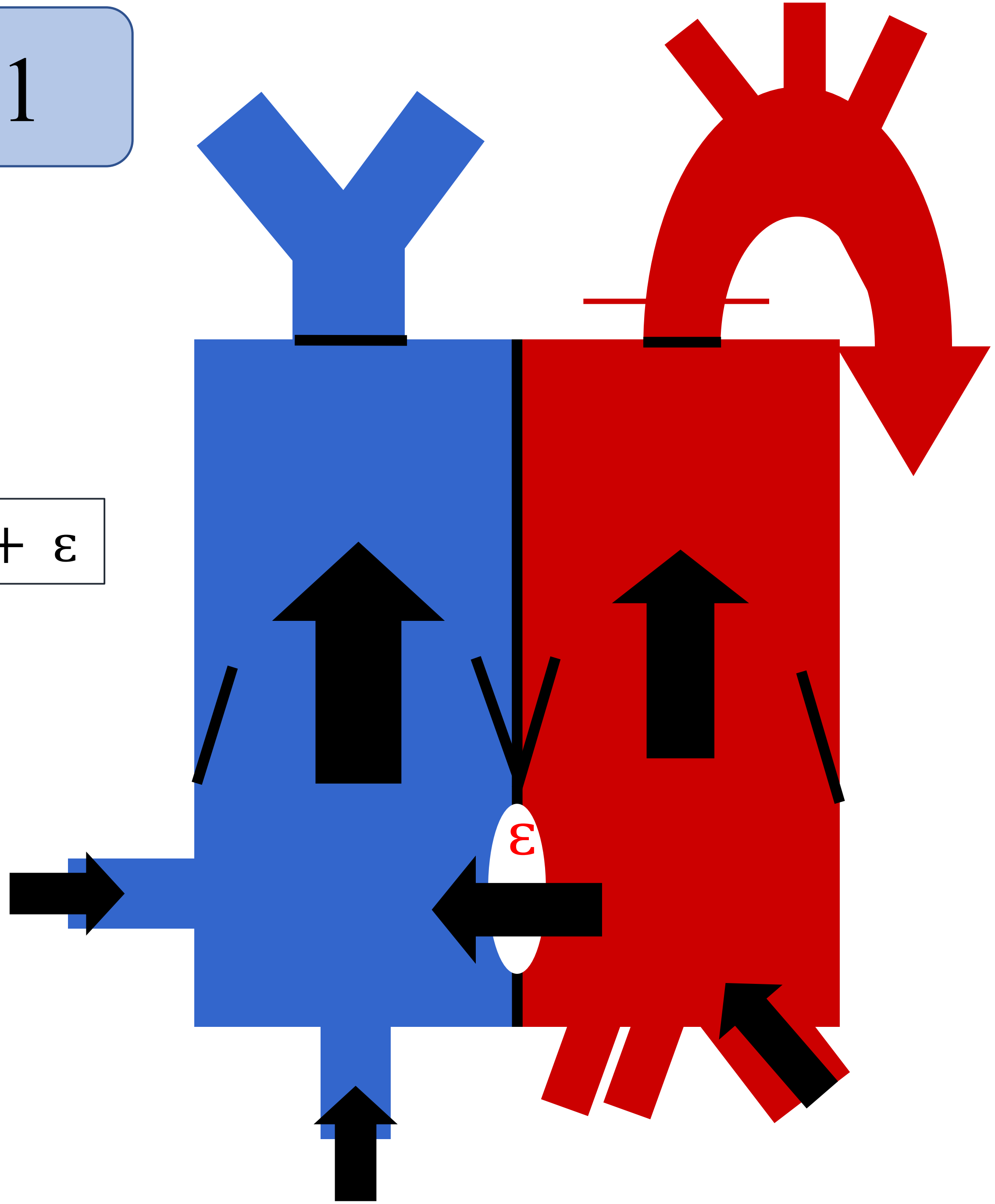
Le shunt gauche-droite est dépendant de :

- La **COMPLIANCE** des ventricules droit et gauche **en diastole**.

=> Surcharge volumétrique diastolique du Ventricule droit

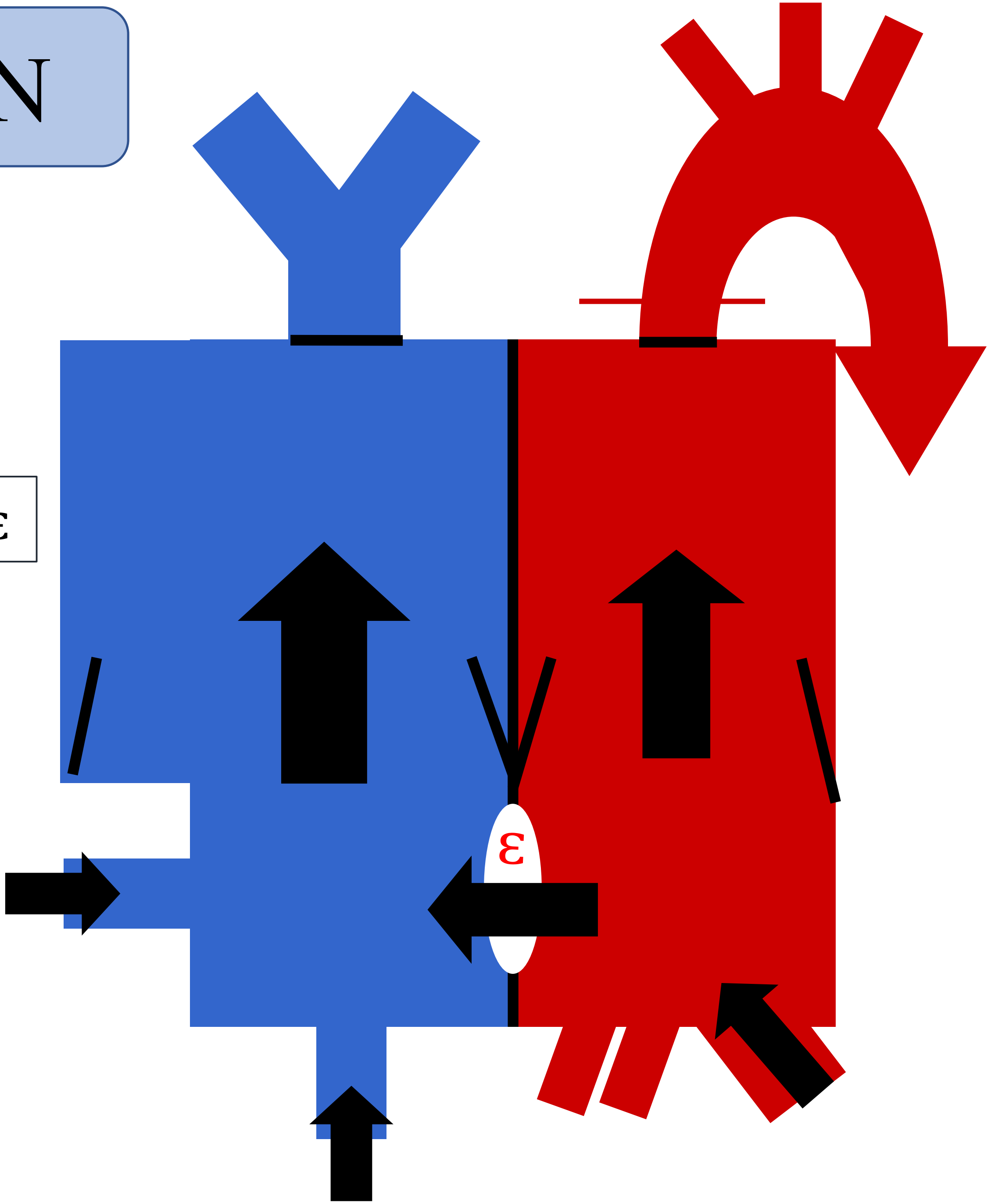
DIASTOLE 1

$$Q(n) = Q(n-1) + \varepsilon$$



DIASTOLE N

$$Q(n) = Q(n-1) + n\varepsilon$$



Evolution

- Une petite CIA peut se fermer spontanément
- Troubles du rythme auriculaire (fibrillation ou flutter) peuvent survenir après 30 ans
- La dysfonction ventriculaire droite est tardive
- L’hypertension artérielle pulmonaire est très rare

Traitement

Indications

- CIA symptomatique
- CIA hémodynamiquement significative, Qp/QS > 1.5 = VD dilaté

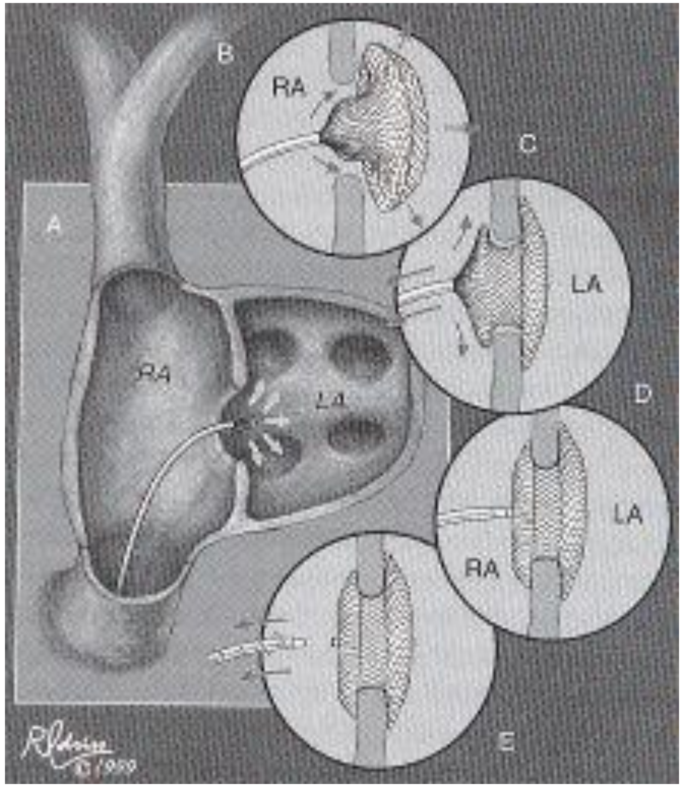
Modalités

- CIA ostium secundum: 90 % de fermeture par KT et 10 % par chirurgie
- Les autres CIA : fermeture chirurgicale

Fermeture per cutanée

Conditions pour fermeture percutanée

- Poids minimum 10 kg
- Anatomie de la CIA type OS
- Berge inférieure au niveau de la VCI
- Le diamètre maximal de la CIA + 14 mm doit être inférieur au diamètre du septum
- Les berges doivent mesurer plus de 5 mm sauf la berge rétro-aortique
- Le septum inter-auriculaire doit être solide



KT vs Chir

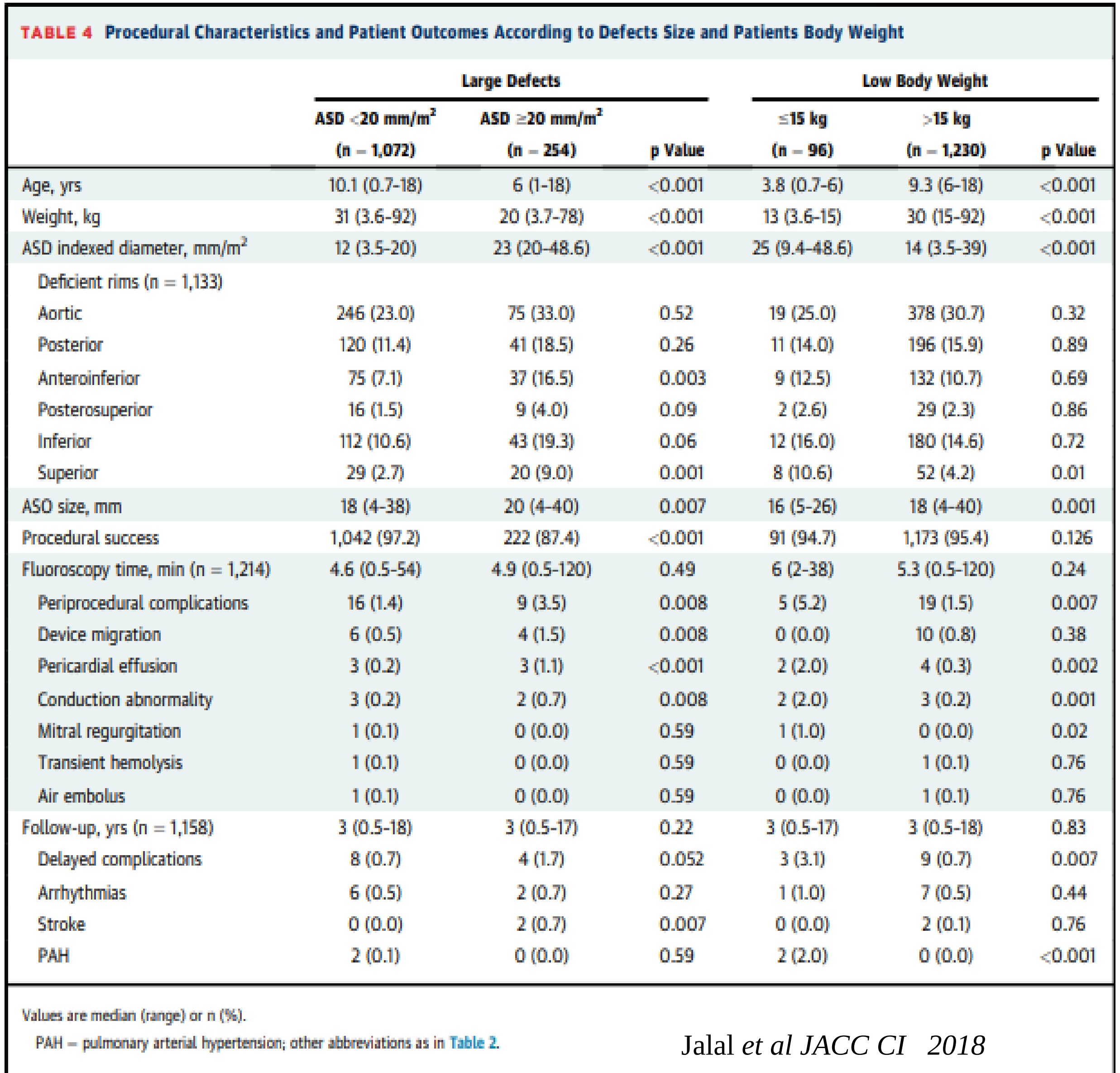


TABLE 4 Procedural Characteristics and Patient Outcomes According to Defects Size and Patients Body Weight

	Large Defects			Low Body Weight		
	ASD $<$20 mm/m^2 (n = 1,072)	ASD $\geq$20 mm/m^2 (n = 254)	p Value	$\leq$15 kg (n = 96)	$>$15 kg (n = 1,230)	p Value
Age, yrs	10.1 (0.7-18)	6 (1-18)	<0.001	3.8 (0.7-6)	9.3 (6-18)	<0.001
Weight, kg	31 (3.6-92)	20 (3.7-78)	<0.001	13 (3.6-15)	30 (15-92)	<0.001
ASD indexed diameter, mm/m^2	12 (3.5-20)	23 (20-48.6)	<0.001	25 (9.4-48.6)	14 (3.5-39)	<0.001
Deficient rims (n = 1,133)						
Aortic	246 (23.0)	75 (33.0)	0.52	19 (25.0)	378 (30.7)	0.32
Posterior	120 (11.4)	41 (18.5)	0.26	11 (14.0)	196 (15.9)	0.89
Anteroinferior	75 (7.1)	37 (16.5)	0.003	9 (12.5)	132 (10.7)	0.69
Posterosuperior	16 (1.5)	9 (4.0)	0.09	2 (2.6)	29 (2.3)	0.86
Inferior	112 (10.6)	43 (19.3)	0.06	12 (16.0)	180 (14.6)	0.72
Superior	29 (2.7)	20 (9.0)	0.001	8 (10.6)	52 (4.2)	0.01
ASO size, mm	18 (4-38)	20 (4-40)	0.007	16 (5-26)	18 (4-40)	0.001
Procedural success	1,042 (97.2)	222 (87.4)	<0.001	91 (94.7)	1,173 (95.4)	0.126
Fluoroscopy time, min (n = 1,214)	4.6 (0.5-54)	4.9 (0.5-120)	0.49	6 (2-38)	5.3 (0.5-120)	0.24
Periprocedural complications	16 (1.4)	9 (3.5)	0.008	5 (5.2)	19 (1.5)	0.007
Device migration	6 (0.5)	4 (1.5)	0.008	0 (0.0)	10 (0.8)	0.38
Pericardial effusion	3 (0.2)	3 (1.1)	<0.001	2 (2.0)	4 (0.3)	0.002
Conduction abnormality	3 (0.2)	2 (0.7)	0.008	2 (2.0)	3 (0.2)	0.001
Mitral regurgitation	1 (0.1)	0 (0.0)	0.59	1 (1.0)	0 (0.0)	0.02
Transient hemolysis	1 (0.1)	0 (0.0)	0.59	0 (0.0)	1 (0.1)	0.76
Air embolus	1 (0.1)	0 (0.0)	0.59	0 (0.0)	1 (0.1)	0.76
Follow-up, yrs (n = 1,158)	3 (0.5-18)	3 (0.5-17)	0.22	3 (0.5-17)	3 (0.5-18)	0.83
Delayed complications	8 (0.7)	4 (1.7)	0.052	3 (3.1)	9 (0.7)	0.007
Arrhythmias	6 (0.5)	2 (0.7)	0.27	1 (1.0)	7 (0.5)	0.44
Stroke	0 (0.0)	2 (0.7)	0.007	0 (0.0)	2 (0.1)	0.76
PAH	2 (0.1)	0 (0.0)	0.59	2 (2.0)	0 (0.0)	<0.001

Values are median (range) or n (%).

PAH = pulmonary arterial hypertension; other abbreviations as in Table 2.

Jalal *et al JACC CI 2018*

Comparison Between Transcatheter and Surgical Closure of Secundum Atrial Septal Defect in Children and Adults Results of a Multicenter **Nonrandomized Trial**

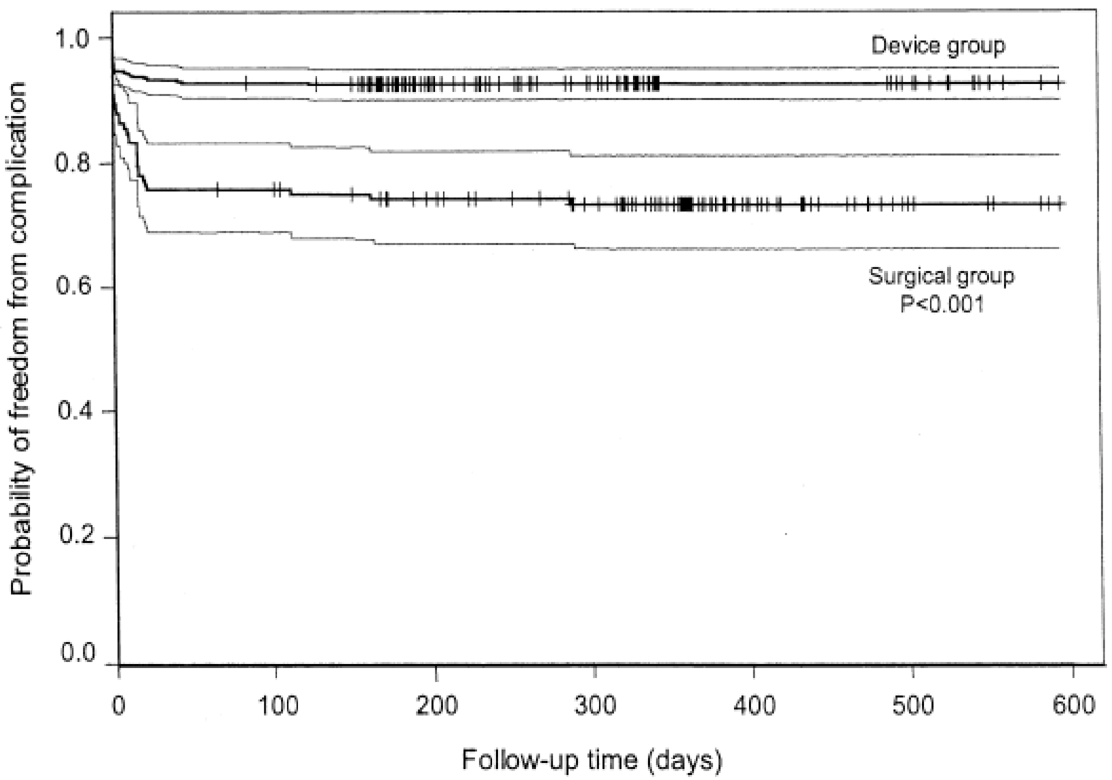


Figure 2. Comparison of curves of freedom from complication probability between device group and surgical group. **Lighter lines** beside each curve are the 95% confidence interval levels.

Du ZD et al J Am Coll Cardiol. 2002

Fermeture chirurgicale

Traitement chirurgical indications :
- CIA os impossibilité ou échec de fermeture percutanée
- CIA sinus venosus avec shunt significatif
- CIA ostium primum avec shunt significatif et/ou fuite significative de la valve AV gauche

Les voies d'abord sont :
- la sternotomie le plus souvent ou mini-sterno
- la thoracotomie postérieure droite chez la jeune fille
- la thoracotomie sous-mammaire droite chez l'adolescente