Pectus Excavatum: Chirurgicus et Pneumologum

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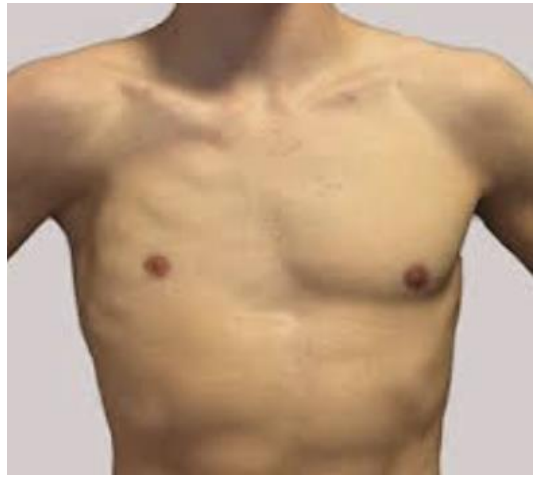
Déformations de la cage thoracique

Pectus excavatum

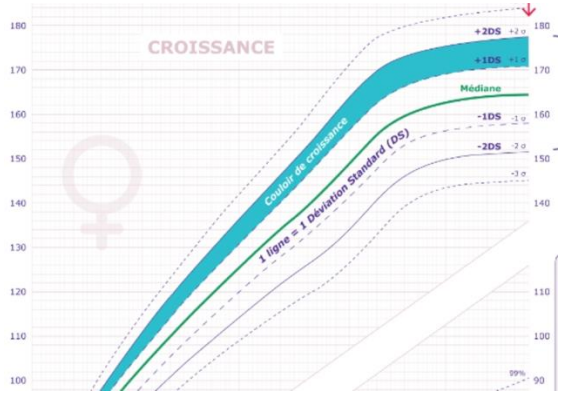
Pectus carinatum

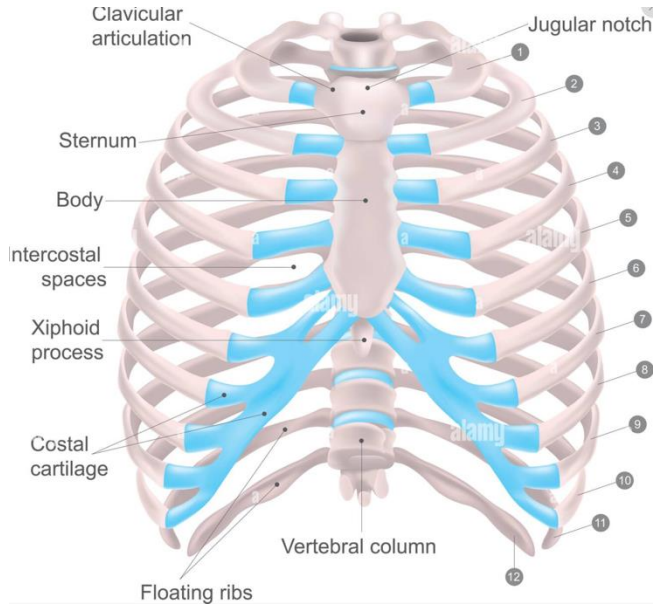
Sd de Poland



Pectus excavatum

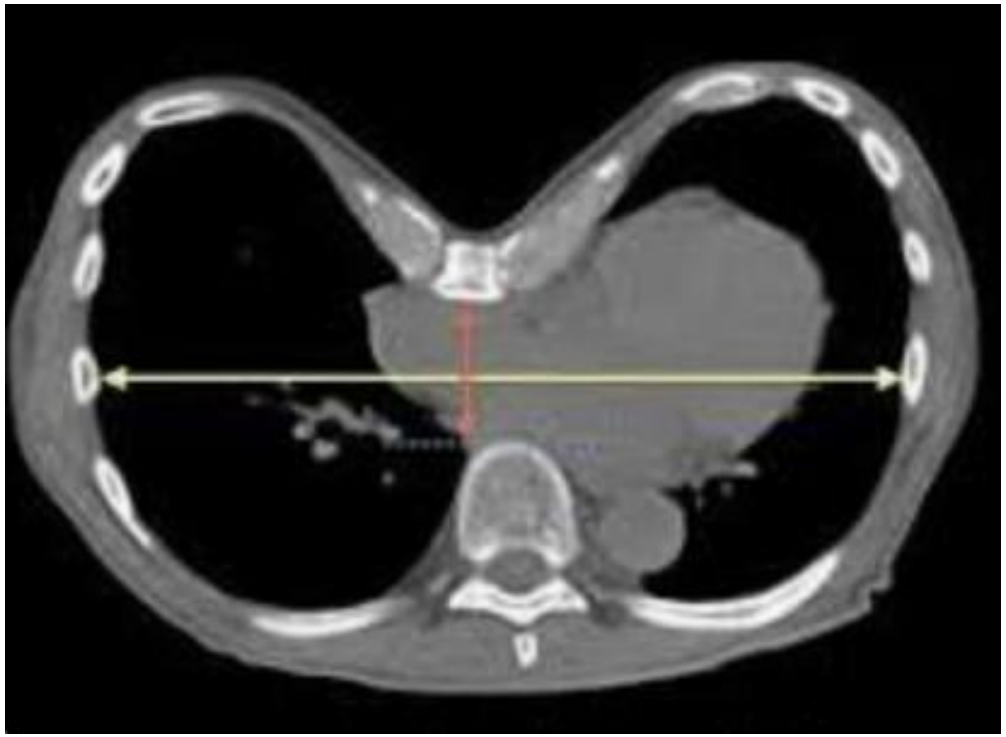
- La + fréquente : 8/1000 naissances
- Homme : 3-5 : 1
- Causes pas claires :
  - Croissance et structure défectueuse des cartilages
  - Mauvaise fusion des côtes avec le sternum au cours du développement embryonnaire.
  - Croissance anormale des cotes
- Pas de régression jusqu'à 6 ans
- N'évolue pas dans 2/3 cas après 12 ans





Pectus excavatum : rarement isolé

- Association fréquente avec une scoliose
- Association
  - Maladie du tissu conjonctif : Marfan , Ehlers-Danlos
  - Maladie neuromusculaire
  - Sd de Noonan , Sd de turner
- Part génétique possible
  - 54 % : ascendant
- Sévérité variable : indice de Haller
  - Normal : 2,5
  - Sévère si > 3,25



Caractéristiques des PE

Evaluations	PE ($n=90$)	HC ($n=90$)	PE versus HC
Posture			
Forward head	74 (82.22%)	64 (71.11%)	**< 0.001**
Rounded shoulder	50 (55.55%)	20 (22.22%)	**< 0.001**
Shoulder height difference	26 (28.89%)	30 (33.33%)	0.06
Kyphosis	19 (31.66%)	12 (13.33%)	**< 0.001**
Scoliosis	5 (5.56%)	2 (2.22%)	**< 0.001**
NYPR	48.10 ± 9.67	59.10 ± 5.91	**< 0.001**
Grip strength			
Dominant side (kg)	51.90 ± 23.40	64.10 ± 22.20	**0.02**[a]
Non-dominant side (kg)	45.30 ± 23.90	58.00 ± 22.40	**0.01**[a]
Sit-reach test (Flexibility, cm)	17.00 ± 7.17	21.160 ± 8.77	**< 0.001**[a]
Sit-up test	21.70 ± 23.90	32.00 ± 5.30	**< 0.001**[a]
Body and back muscle strength (kg)	16.10 ± 5.96	32.70 ± 15.20	**< 0.001**[a]
Physical activity			
Low	16 (17.78%)	9 (10.00%)	**< 0.001**
Moderate	50 (55.55%)	55 (61.11%)	
High	8 (8.89%)	26 (28.89%)	
Sitting (min)	411 ± 174.90	424 ± 96.60	0.89[a]
Total score (MET)	1734 ± 1462	3050 ± 2388	**< 0.001**[a]

Evaluations	PE	HC	PE versus HC
Brief symptom inventory			
Depression	11.72 ± 10.63	9.01 ± 8.60	0.26
Anxiety	12.00 ± 9.78	8.22 ± 7.28	**0.05**
Somatization	6.94 ± 5.53	4.42 ± 4.34	**0.02**
Negative self	9.34 ± 5.53	6.65 ± 5.90	0.11
Hostility	8.06 ± 5.75	5.98 ± 4.06	**0.06**
Body image scale	141 ± 29.30	149 ± 28.00	0.21
Social Appearance Anxiety Scale	30.01 ± 12.30	32.80 ± 15.30	0.51
Social Anxiety Scale for Adolescents			
Fear of negative Evaluation	12.94 ± 7.01	11.36 ± 5.68	0.48
Social avoidance and distress-general	10.65 ± 4.41	10.29 ± 5.14	0.85
Social avoidance and distress-new	8.65 ± 2.11	6.26 ± 2.37	**< 0.001**
Pediatric quality of life inventory			
The total score of the Pediatric Quality of Life Inventory scale	72.27 ± 20.57	88.75 ± 5.93	**0.03**
Total physical health score	70.84 ± 21.17	85.50 ± 6.71	**0.05**
Total psychosocial health score	72.01 ± 18.90	85.98 ± 5.48	**0.04**

N.Alaca . Pediatric Surgery International (2021) 37:765–775Comparison of physical functions and psychosocial conditions between adolescents with pectus excavatum, pectus carinatum and healthy controls

Plaintes des patients

Plaintes cosmétiques

- Enfant
- Adolescent
- Femme

Plaintes fonctionnelles

- Intolérance à l'exercice
- Dyspnée
- Douleur thoracique
- Absence d'endurance

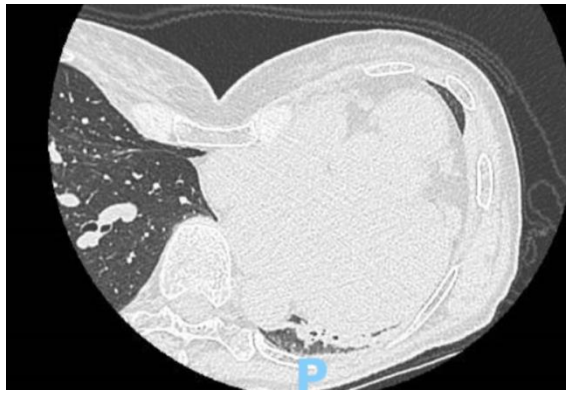
Aucune corrélation avec la sévérité du PE

Pectus : FdR d' infections pulmonaires ?

- Morphotype identique aux patients avec MNT
- Association MNT et PEX

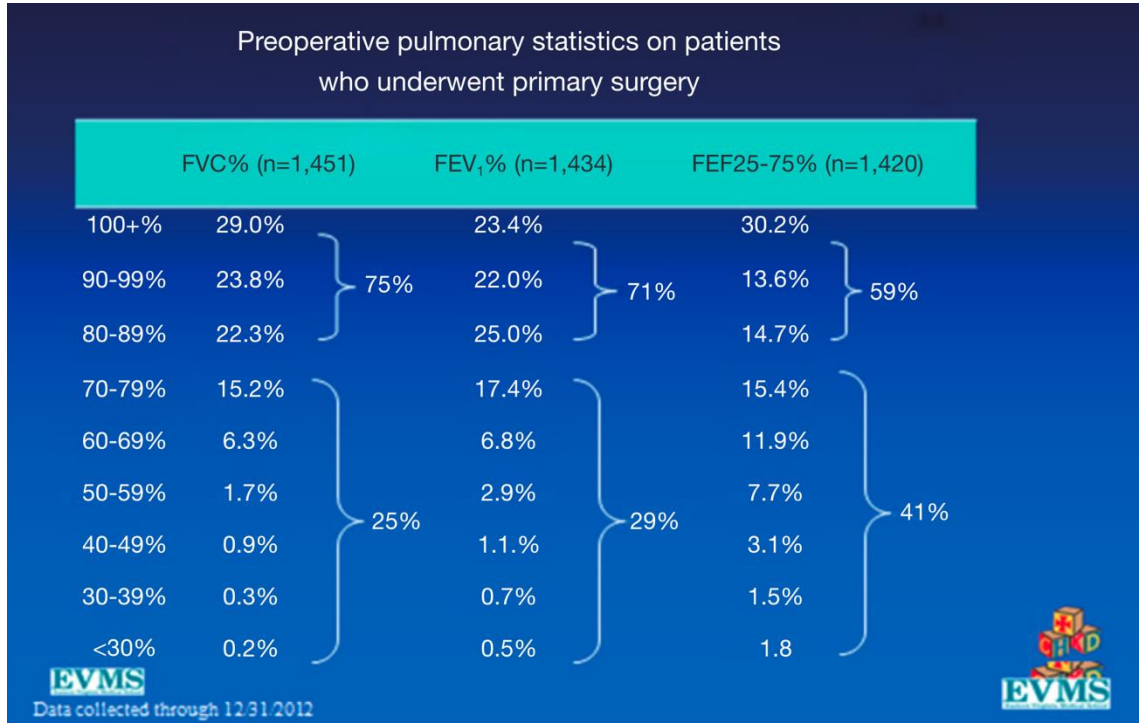
Variable	Control Subjects ($n = 101$)*	Patients with pNTM ($n = 103$)*	*P* Value
Height, cm	161.83 (8.40)	166.94 (7.62)	<0.0001
Weight, kg	63.12 (1.49)	61.77 (1.23)	0.48
BMI, kg/m^2	23.98 (5.07)	22.06 (3.81)	0.003
Percent body fat, %	31.28 (7.60)	28.46 (7.42)	0.008
Total body fat, kg	20.43 (8.82)	17.84 (6.61)	0.02
Scoliosis, N (%)†	10 (13.3)	32 (31.1)	0.006
PEX, N (%)‡	13 (65)	90 (87)	0.01

- Association Pectus et dilatation des bronches localisée
  - Lingula



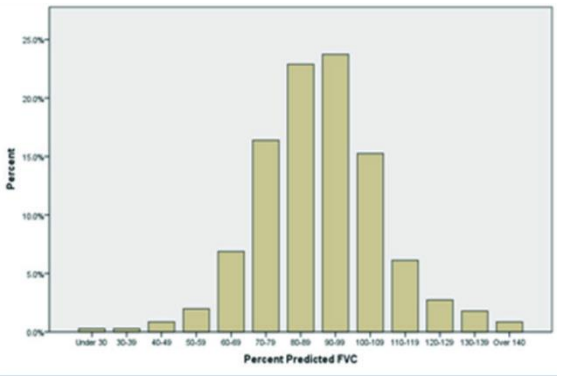
Mkartalija – AJRCCM 2013 - Patients with Nontuberculous Mycobacterial Lung Disease Exhibit Unique Body and Immune Phenotypes

Fonction respiratoire : peu d'impact



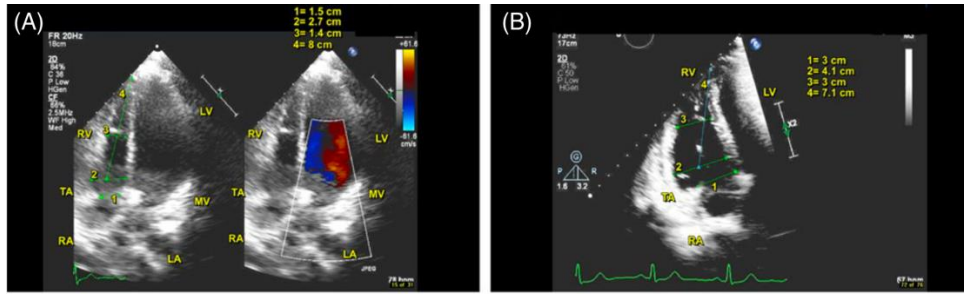
	Avant la chirugie	Après la chirurgie
CV	87 %	92,6 %
VEMS	86 %	90 %
CPT	94 %	100 %

// Indice de Haller (> 5)

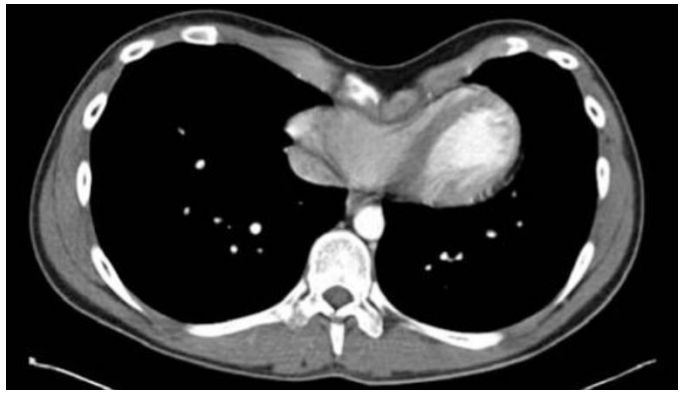


Kelly et al. Ann Cardiothorac Surg 2016;5(5):466-475 Pulmonary function in pectus excavatum

Fonction cardiaque



- Evaluation de la compression des cavités droites
  - OD
  - VD
  - Fonction : trouble de remplissage du VD
- Evaluation de la valve mitrale
  - Distorsion mécanique de la valve mitrale → prolapsus de la valve mitrale
  - 40 % (vs 4-8 % population générale)



Jaroszewski DE et al. Right ventricular compression observed in echocardiography from pectus excavatum deformity. J Cardiovasc Ultrasound. 2011;19:192–195.
AY Salama . Echocardiography. 2019;36:15 0 –16 3 . Incremental value of three-dimensional transthoracic echocardiography over the two-dimensional modality in the assessment of right heart compression and dysfunction produced by pectus excavatum
A.Sanaglioni Diagnostics 2024, 14, 2488. Prevalence of Mitral Valve Prolapse Among Individuals with Pectus Excavatum: A Systematic Review and Meta-Analysis

Altération de la capacité à l'exercice

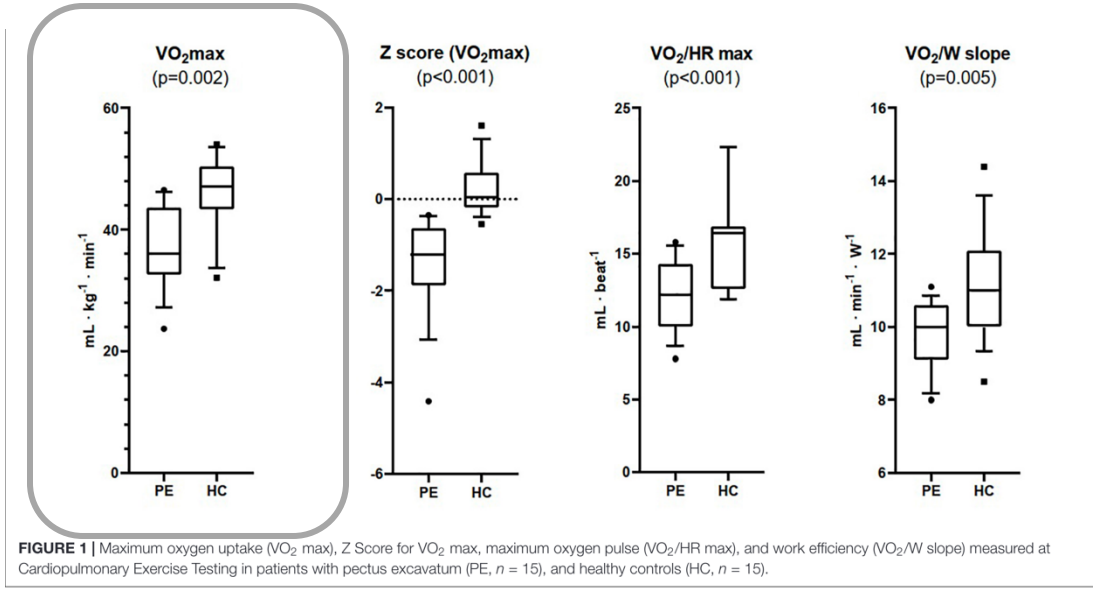


FIGURE 1 | Maximum oxygen uptake (VO_2 max), Z Score for VO_2 max, maximum oxygen pulse (VO_2/HR max), and work efficiency (VO_2/W slope) measured at Cardiopulmonary Exercise Testing in patients with pectus excavatum (PE, n = 15), and healthy controls (HC, n = 15).

- Etude francaise : 60 patients – 26 ans
- Limitation à l'augmentation du V_T (n = 18) (41 ± 5 % de la CVF au pic versus 51 ± 1,4 %)
- Elévation du gradient alvéolo-artériel en oxygène au pic de l'exercice (n = 5) (47 ± 23 mmHg versus 20 ± 7,5 mmHg), associée à un foramen ovale perméable
- Limitation cardiovasculaire: pouls d'oxygène bas au pic (57 ± 9 % versus 90 ± 20 %)

L.Casatori . Frontiers in physiology . Differential Influence of Physical Activity on Cardiopulmonary Performance and Stroke Volume Assessed at Cardiopulmonary Exercise Test in Pectus Excavatum: A Pilot Study

Cavestri Revue de maladies respiratoires 2010. Exploration fonctionnelle à l'exercice des patients présentant un pectus excavatum

Altération de la Capacité à l'exercice

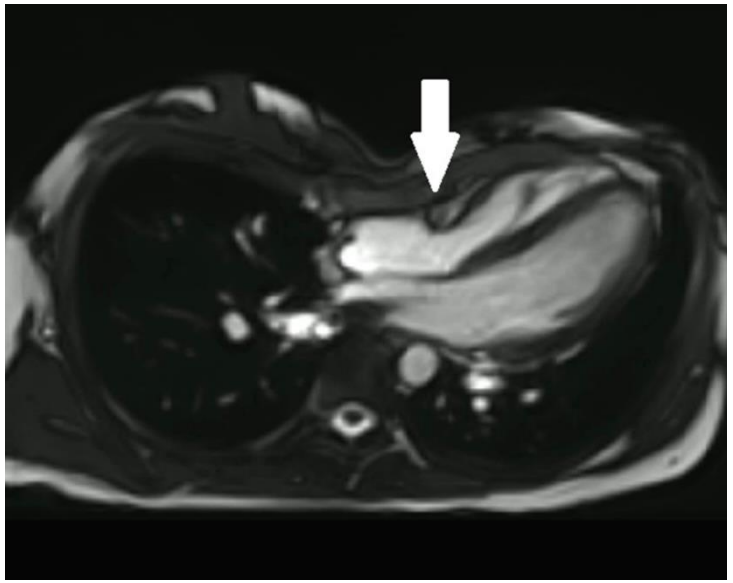
	Total **n=70**	Group A **n=33**	Group B **n=37**	**Comparison of group AandB**
Age, years; mean±SD	20.8 (6.6)	20.3 (5.0)	21.4 (7.8)	*0.51*
M/F	60/10	31/2	29/8	*0.09*
Height, cm; mean±SD	177.7 (8.3)	178.8 (7.7)	176.7 (8.8)	*0.28*
Weight, kg; mean±SD	64.4 (12.1)	64.8 (10.2)	64.1 (13.8)	*0.80*
Haller's Index; mean±SD	4.1 (1.4)	4.2 (1.3)	4.0 (1.6)	*0.56*
Spirometry				
FEV_1, % predicted; mean±SD	92.0 (12.8)	94.3 (12.8)	90.7 (12.7)	*0.25*
FVC, % predicted; mean±SD	91.8 (11.9)	93.9 (12.1)	89.9 (11.5)	*0.16*
IC, % predicted; mean±SD	86.0 (14.8)	87.5 (13.0)	84.6 (16.3)	*0.43*

Cardiopulmonary exercise data (**Mean±SD)**	Total group **n=70**	Group A **n=33**	Group B **n=37**	**Comparison of group A and B**
Δ HCO_3^- (peak – rest)	−7.1 (2.5)	−8.0 (2.6)	−6.2 (2.2)	*0.003*
Δ lactate	8.4 (2.8)	9.7 (2.7)	7.4 (2.4)	*0.002*
VO_2 max, % predicted	78.0 (13.7)	89.6 (7.8)	67.7 (8.6)	*<0.0001*
Work, % predicted	87.9 (14.2)	97.7 (10.1)	79.2 (11.3)	*<0.0001*
VO_2/work	9.51 (1.3)	10.03 (0.16)	9.04 (0.23)	*0.0012*
AT, % predicted	43.5 (10.0)	49.7 (9.1)	38.1 (7.8)	*<0.0001*
O_2 pulse, % predicted	88.9 (16.3)	101.8 (11.7)	77.4 (9.8)	*<0.0001*
BR%	49.9 (13.2)	44.2 (10.8)	54.9 (13.1)	*0.0002*

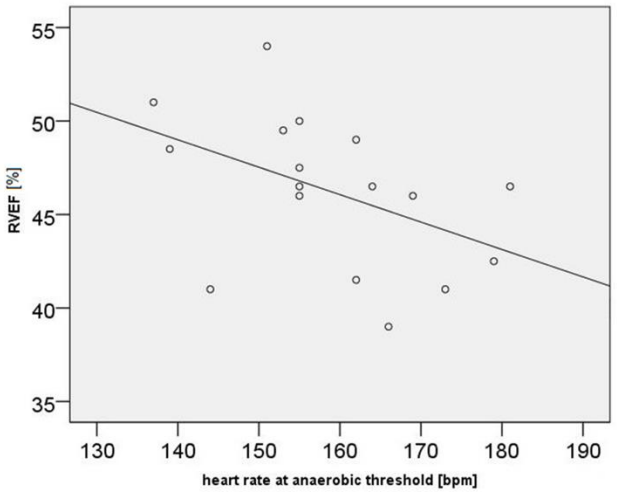
- Indépendante de la sévérité du PE
- Pas de limitation ventilatoire
- Limitation périphérique
  - Déconditionnement
- Limitation cardiovasculaire

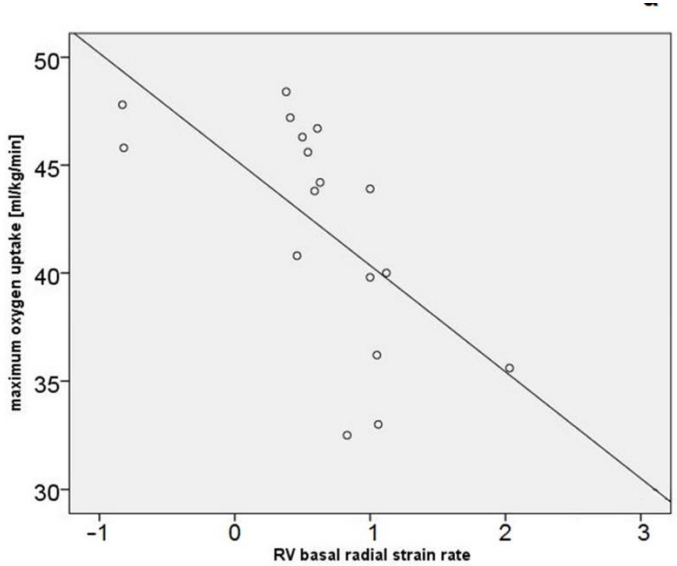
Satur CMR, et al. BMJ Open Resp Res 2021;8:

Dysfonction diastolique a l’effort



- Compression du VD par le sternum
- Trouble de remplissage du VD
- Augmentation insuffisante du VES a l’effort
- Compensation par une augmentation de la FC





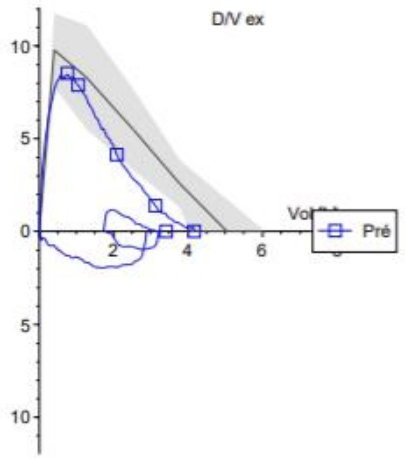
A.Lollert .Diagnostics 2024, 14, 2758. Cardiac Magnetic Resonance Imaging with Myocardial Strain Assessment Correlates with Cardiopulmonary Exercise Testing in Patients with Pectus Excavatum

La chirurgie améliore la capacité à l'exercice

Variable	Preoperative	Postoperative	*P* value
FVC (L)	4.7 ± 0.7 (91% ± 4%)	4.5 ± 0.8 (90% ± 4%)	.40
FEV_1 (L)	3.9 ± 0.8 (94% ± 2%)	3.9 ± 0.9 (94% ± 2%)	.90
TLV (L)	6.8 ± 1.2 (95% ± 3%)	6.7 ± 1.4 (95% ± 3%)	.81
PImax (cm H_2O)	79 ± 21 (76% ± 16%)	90 ± 17 (88% ± 10%)	<.01
SNIP (cm H_2O)	69 ± 19 (70% ± 13%)	77 ± 18 (78% ± 14%)	<.01
VO_2 (mL/kg/min)	30.8 ± 6.9 (73% ± 10%)	34.4 ± 8.6 (82% ± 11%)	<.01
RER (VCO_2/VO_2)	1.22 ± 0.14	1.23 ± 0.11	.80
HR (beats/min)	178 ± 10 (93% ± 4%)	171 ± 12 (91% ± 6%)	.06
Oxygen pulse (mL/beat)	11.5 ± 3.2 (80% ± 12%)	12.9 ± 3.5 (91% ± 14%)	<.05

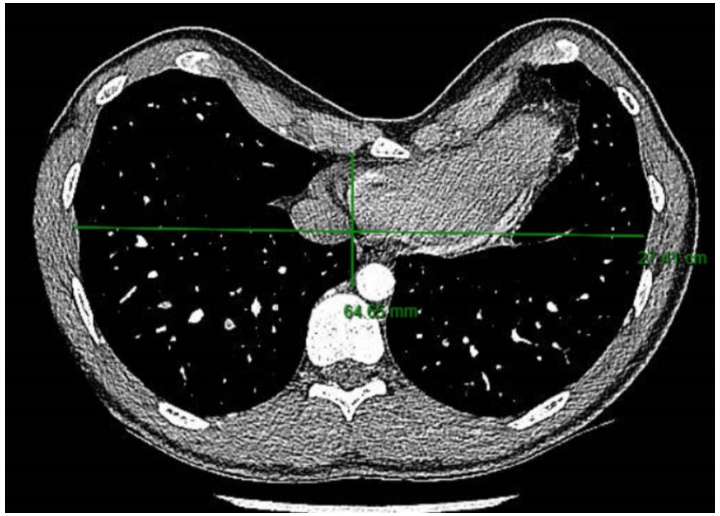
R.Neviere . JTCVS 2013;145:605-6 .Pectus excavatum repair improves respiratory pump efficacy and cardiovascular function at exercise

Mr B – 22 ans – Dyspnée d'effort



	Théo	Pré	%th
Pos		Assis	
SPIROMETRIE			
CVF	5.03	4.14	82
VEMS	4.27	3.41	80
VEMS%CF	82.71	82.27	99
VEMS%CV	82.71	82.27	99
DEM 25/75	5.00	3.48	70
DEM75	8.31	7.89	95
DEM50	5.47	4.14	76
DEM25	2.55	1.39	55
DEP	9.76	8.50	87
VIMS		1.87	
DIM50		1.89	
RESISTANCES			
CV Max	5.26	4.07	77
CPT	6.82	5.86	86
VR	1.60	1.79	112
VR%CPT	23.71	30.51	129
VRE	1.61	1.51	94
CRFpl	3.21	3.29	103
RAW	3.06	3.11	102
G AW	0.33	0.32	98
SR AW	12.00	11.66	97
SG AW	0.08	0.09	103
DLCOcSB	34.79	36.89	106
KCOc	5.10	7.09	139
Hb		14.60	
VA_SB	6.67	5.21	78

GAZ DU SANG



Indice de Haller : 5,5

Echographie cardiaque normale

Dépense Métabolique

Résumé		Repos	SV1 [Manuel]	SV1%théo [Manuel]	PIC VO2	PIC VO2 %Th	Théo Max
Watt	[W]	0	106	44	188	78	240
V'O2	[mL/min]	204	1279	43	2167	72	2994
V'O2/kg	[(mL/min)/kg]	3.2	20.0	43	33.9	72	46.8
V'CO2	[mL/min]	181	1196	-	2594	-	-
QR		0.89	0.94	-	1.20	-	-
SpO2	[%]	100	97	-	95	-	-
Dys Fat		-	-	-	-	-	-
RPE Dys		-	-	-	-	-	-

Paramètres Ventilatoires

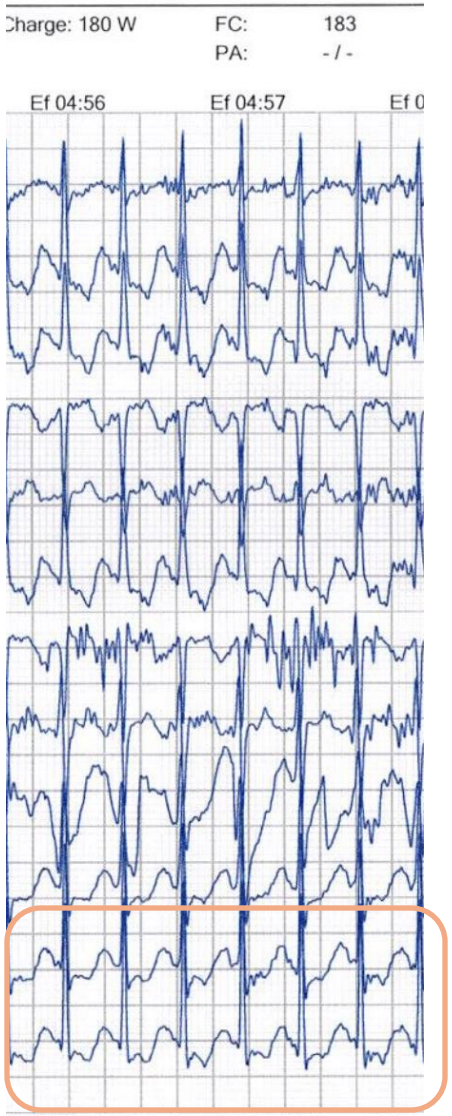
V'E	[L/min]	6	31	26*	77	65*	119*
VTex	[L]	0.605	1.661	-	2.068	-	-
FR	[1/MIN]	10.5	18.4	44	37.4	90	41.6
EqO2		25.6	22.4	-	33.8	-	-
EqCO2		28.9	23.9	-	28.2	-	-
VDe/VT	[%]	11	13	79	14	88	16
RR VEMS%	[%]	95	74	265	35	126	28

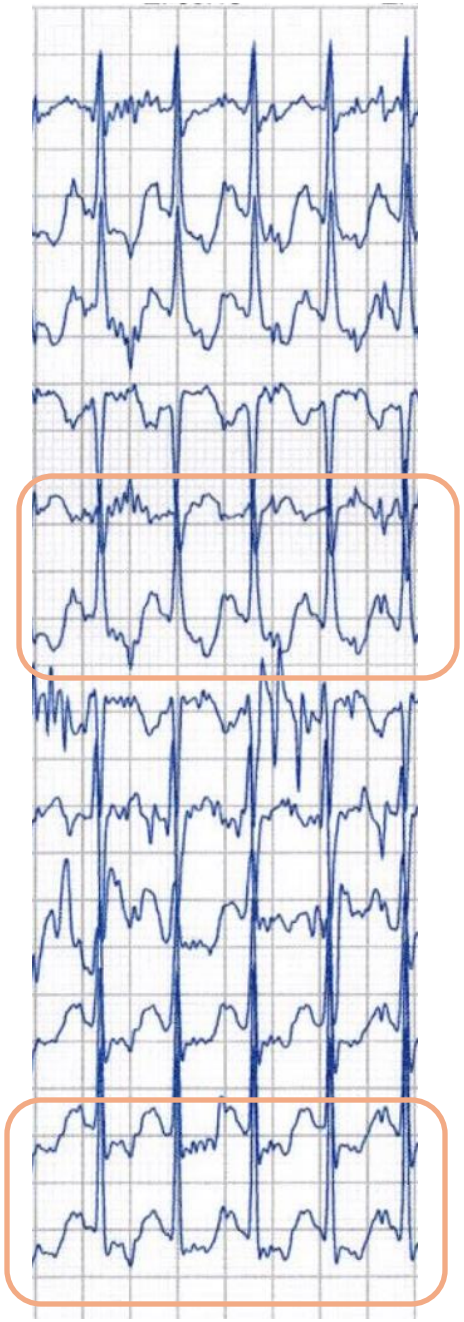
Paramètres Cardio-vasculaires

FC	[1/MIN]	70	144	73	187	94	198
PoulsO2	[mL]	2.9	8.9	59	11.6	77	15.1
Psys	[mmHg]	120	134	-	156	-	-
Pdia	[mmHg]	58	69	-	77	-	-

Hématose

Temps	[min]	01:18	08:19	
PO2 art.	[mmHg]	82.60	119.00	
PCO2 Art.	[mmHg]	39.90	31.00	
pHa		7.44	7.43	
Hb	[g(Hb)/dL]	-	-	
Azote U.	[g/day]	15.00	15.00	
SaO2	[%]	97	98	
O2	[L/min]	-	-	
VDf/VT	[%]	13	-	
AaDO2	[mmHg]	15.83	1.37	





IRM cardiaque :

Pectus excavatum sévère avec un index de Haller à 5,9. Aplatissement consécutif des cavités cardiaques droites, notamment de la paroi libre du ventricule droit, avec une dilatation modérée de l'oreillette droite. Petit épanchement péricardique réactionnel en regard.

Comment prendre en charge les Pectus excavatum ?

Bilan de retentissement fonctionnel

- EFR
- TDM thorax injecté
- ECG - ETT
- IRM cardiaque
- EFX

We recommend that all patients with severe pectus excavatum and exercise intolerance undergo a cardiopulmonary exercise test because it is the best test to identify exercise intolerance that is caused by pectus excavatum. A ventilatory update of oxygen (VO_2) max below 85% of predicted is regarded as an abnormality.
(Class I, Level of Evidence B)

Jansen N . Ann Thorac Surg 2023;116:191-9Pectus Excavatum: Consensus and Controversies in Clinical Practice
The pectus care guidelines: best practice consensus guidelines from the joint specialist societies SCTS/MF/CWIG/BOA/BAPS for the treatment of patients with pectus abnormalities - European Journal of Cardio-Thoracic Surgery 2024

Qui doit prendre en charge les Pectus excavatum ?

Pneumologue/ cardiologue/ chirurgien thoracique pour le bilan

Si limitation à l'exercice

Retentissement cardiaque

↓

Chirurgie thoracique

Si gène esthétique

↓

Chirurgie plastique