Very unusual clinical-electrocardiographic picture in a young adult Caucasian male

A 20-year-old Caucasian man was admitted to the emergency department with abdominal pain and worsening dyspnea, with 6 months of symptom evolution. He was class I or II NYHA, with no history of chest pain, palpitations, or syncope. He had no relevant medical history and an unknown family history because he was adopted.

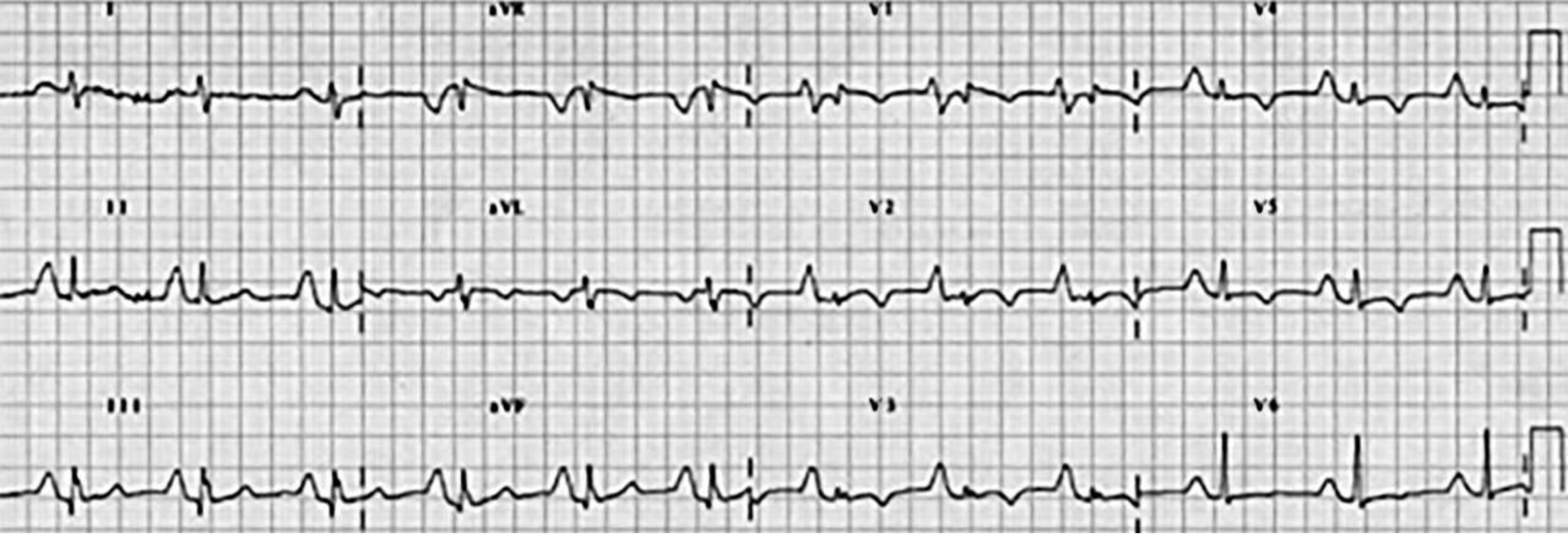
Physical examination revealed tachypnea with peripheral cyanosis, a pulse rate of 100 beats/min, normal blood pressure, apyrexia, and oxygen saturation of 95% on room air. The patient had jugular venous distension of about 8 cm at 45°, with hepatojugular reflux. Pulmonary auscultation was normal, and a grade VI holosystolic murmur in the left sternal margin was present on cardiac auscultation. The abdomen was diffusely painful, and the liver edge was palpable 5 cm below the right costal margin.

Chest radiography showed cardiomegaly with a normal pulmonary vasculature pattern.

Raza blanca, masculino, 20 años ingresó en la sala de urgencia con queja de dolor abdominal y empeoramiento de la disnea, con 6 meses de evolución de los síntomas. Estaba en clase I o II de NYHA. Sin antecedentes de dolor torácico, palpitaciones ni síncope. No tenía antecedentes médicos relevantes y los familiares eran desconocidos por ser hijo adoptivo.

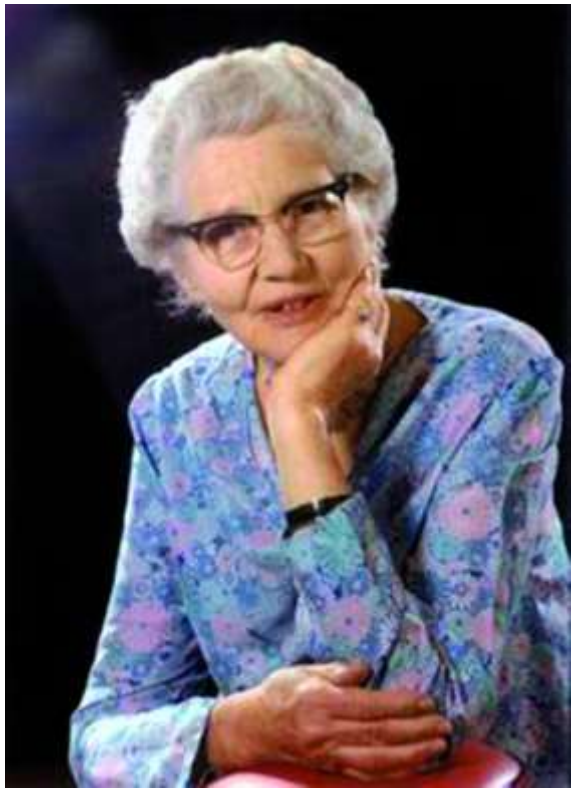
Examen físico: taquipneico, acrocianótico, pulso regular de 100 latidos/min, presión arterial normal, afebril y con saturación de oxígeno de 95% en aire ambiente. En el cuello destacaba distensión yugular venosa de unos 8 cm a 45°, con reflujo hepatoyugular. Auscultación pulmonar normal y la cardíaca con soplo grado 6 holosistólico en el margen esternal izquierdo. Abdomen presentaba un dolor difuso y el borde hepático palpable a 5 cm por debajo del borde costal derecho.

Rx tórax PA mostró cardiomegalia con patrón vascular pulmonar normal.



Sinus rhtytm, HR 56bpm, giant tall and wide P wave: Himalayan P waves: tall (>5 mm) and peaked, most prominent in lead II , SÂP +75 degree, prolonged P-duration (175ms) P-voltage from V1-V4 > QRS amplitude(diminute QRS amplitude from V1 –V4): Modified Peñaloza and Tranchesi sign: QRS complexes of low voltage in V_1 contrasting with QRS complexes of normal voltage or increased in V_2, In the presente case diminute QRS complex form V1 to V3 contrasting with giant P wave SÂT + 90º, T-Wave Inversion (TWI) from V1 to V5 . Modified Peñaloza and Tranchesi sign: QRS complexes of low voltage in V1 contrasting with QRS complexes of normal voltage or increased in V2

Hymalayan P wave denomination



Dr Helen Taussig was a pioneer as a woman in academic medicine and as a leader in pediatric cardiology. She was the second woman promoted to Professor at The Johns Hopkins University School of Medicine, although her promotion and recognition occurred much later than her achievements warranted.

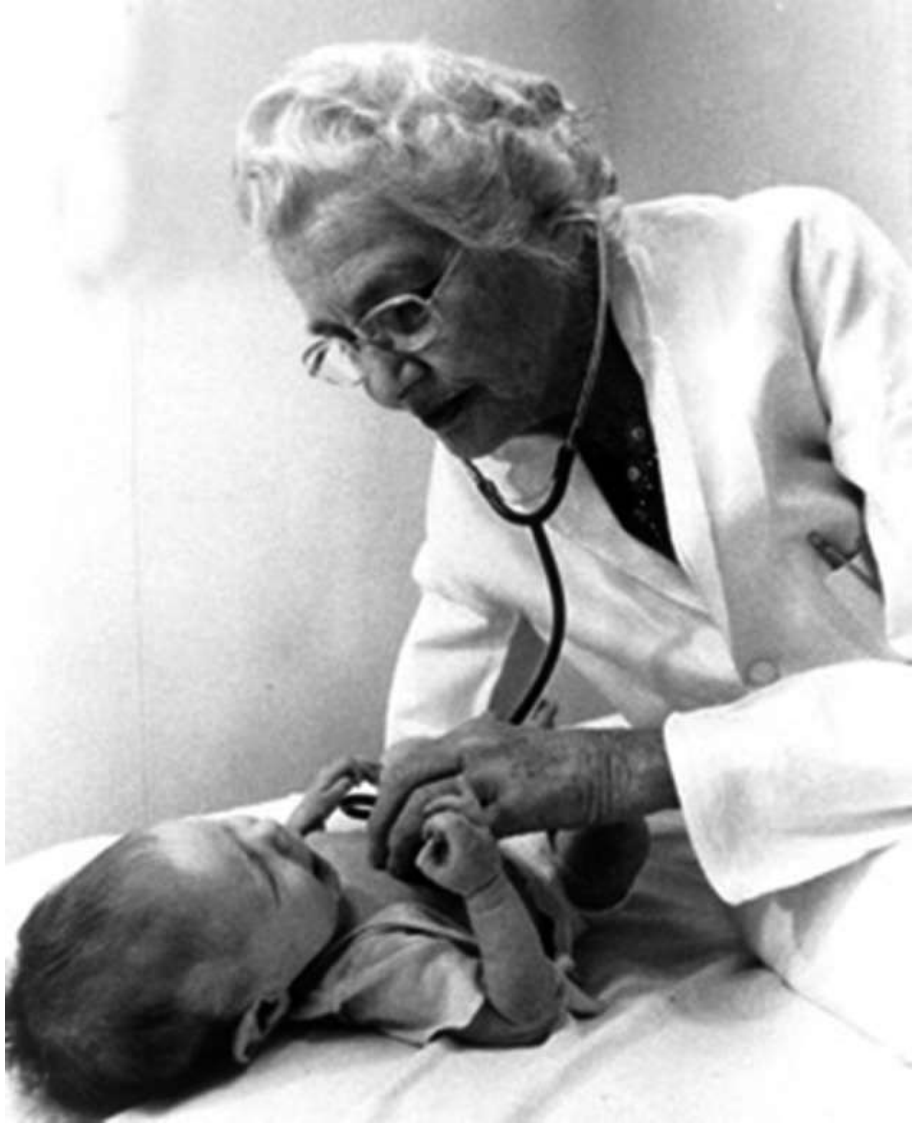
The development of treatment of congenital heart defects was hastened by her careful observations as a clinician and her collaboration with Dr Alfred Blalock and other specialists. Together with Alfred Blalock, Helen Taussig rose to fame because of the Blalock-Taussig shunt. This is a surgical procedure in children who suffer from hypoxia because of a congenital cardiac malformation. Because of this affliction, they are blue. Less known is the fact that dr. Taussig also played a crucial role in the prohibition of thalidomide in the USA, the drug that had been responsible for the Softenon tragedy in Europe.

Dr Taussig received recognition not only for her pioneering work but also for her advocacy, which led to the Food and Drug Administration not permitting use of thalidomide in the United States and her leadership and election to president of the American Heart Association. She received the Presidential Medal of Freedom in 1964

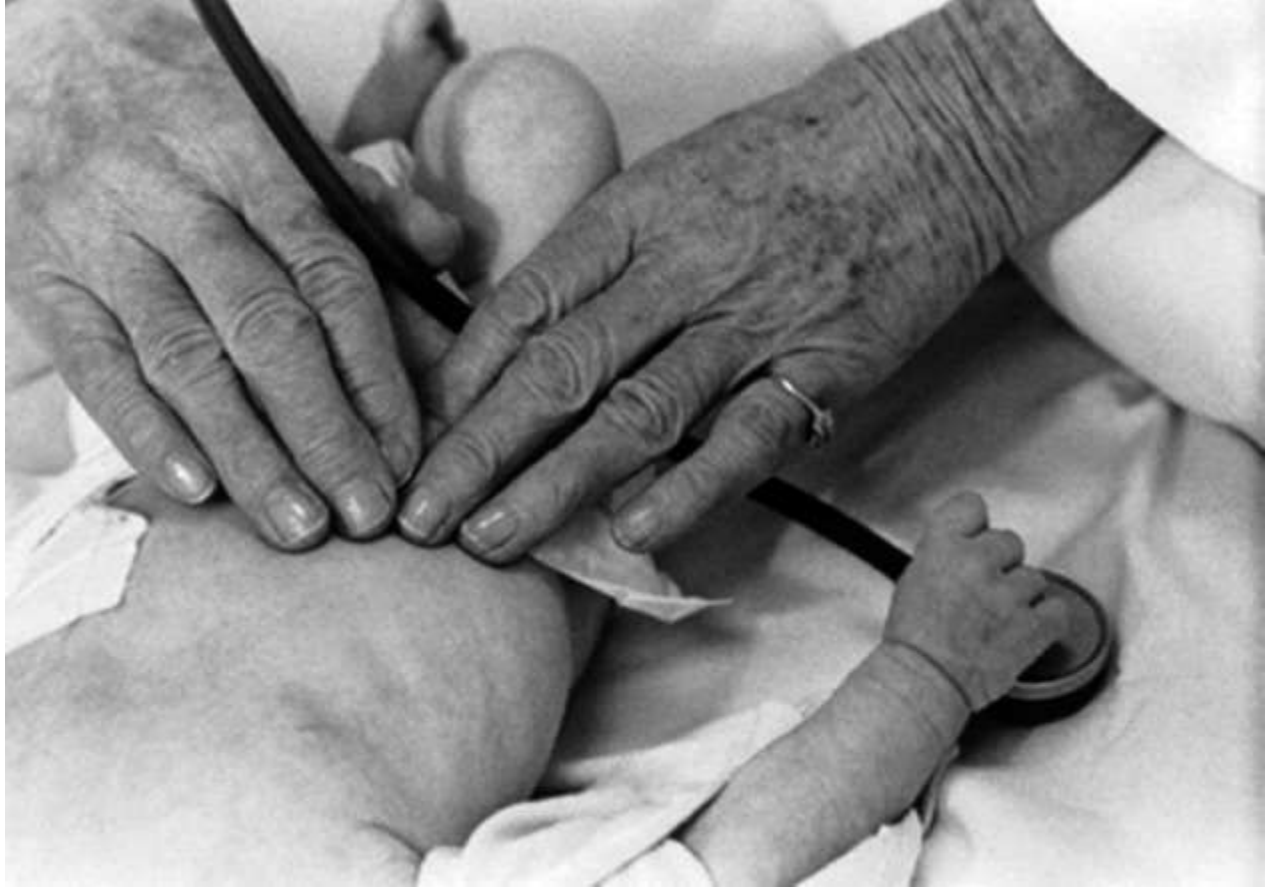
Dyslectic and deaf

Helen Taussig was born on the 24th of May, 1898, in Cambridge, Massachusetts, as the youngest of four children. Her father, Frank Taussig, was a professor in Economy at Harvard University. Her mother had been one of the first female graduates at the Radcliffe College, where she had studied biology and zoology. However, she died of tuberculosis when Helen was only 11 years old. Helen's grandfather, William Taussig, had been a teacher in a school for blind children. As a tribute to his services, a school was named after him: the William Taussig School for Handicapped Children. As Helen herself said, she descended "from a direct line of teachers, and an indirect line of doctors".

In the Cambridge School for Girls, she didn't exactly get top grades. After an innocent flu, she had gotten a middle ear infection, which rendered her half deaf. Furthermore, she was having trouble reading. When her father discovered her dyslexia, he helped her spelling words and reading texts. A successful endeavour, as it turned out, as Helen followed into her mother's footsteps and took up studies at the Radcliffe College in 1917. Here, she attained good grades and even became the college's tennis champion. Two years later, she left for Berkeley University in California, where she got her baccalaureate degree in 1921. Afterwards, she wanted to continue her education at Harvard University, but it wasn't until 1945 that this male bastion allowed women to enter.



Dr Taussig using the stethoscope, before the hearing problem



Dr Taussig using her fingertips to examine her young patients' heart

"By the time Taussig graduated from Hopkins, she had lost her hearing and relied on lip-reading and hearing aids for the rest of her career."

She learned to "listen with her fingers" to her patients' hearts.

ECG analysis and theoretical considerations by

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Right Atrial Enlargement Etiologies

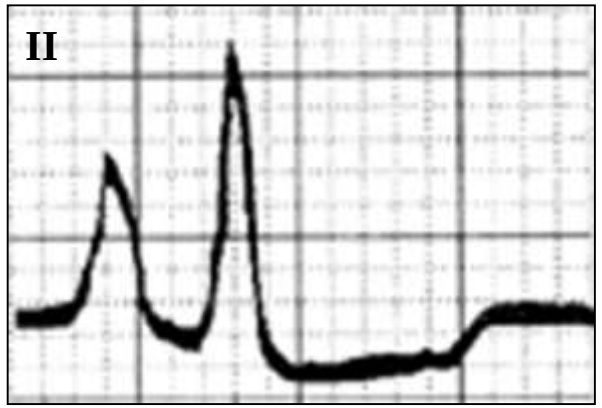
I) Congenital causes: “congenital P”

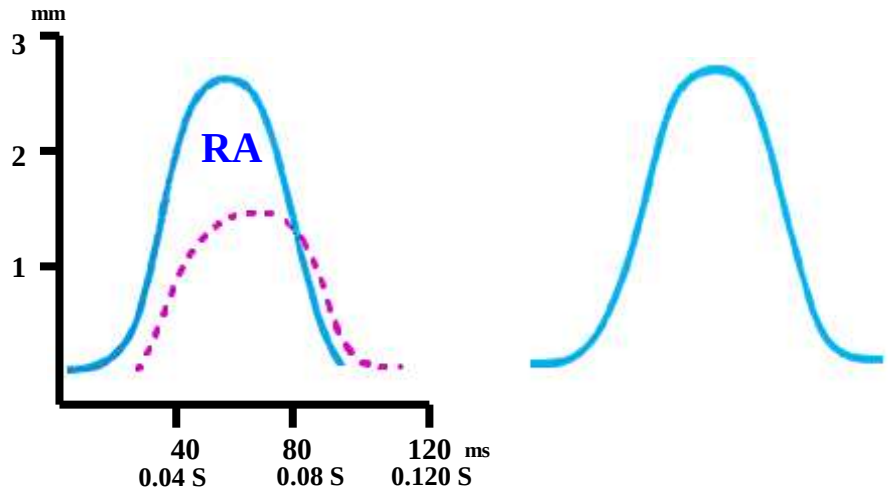
- Ebstein’s anomaly - “Himalayan P waves” of Taussig;
- Uhl's anomaly or parchment heart
- Tricuspid atresia: “P tricuspidale” of Gamboa (10%);
- Moderate pulmonary stenosis “gothic” P (30%);
- Severe pulmonary stenosis P >2.5 mm 75%;
- Eisenmenger Syndrome;
- Atrioventricular septal defect (LV → LA). RAE or BAE 60%;
- Tetralogy of Fallot (T4F): only in 5% there are criteria of RAE.

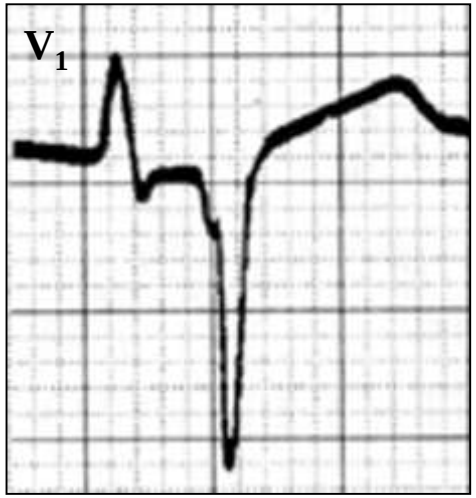
II) Acquired causes

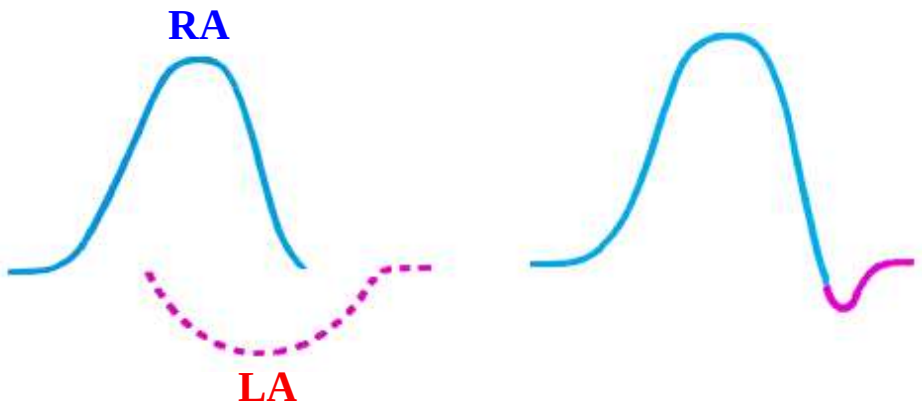
- Cor pulmonale - emphysema - COPD: “P pulmonale”
- Tricuspid stenosis;
- Tricuspid regurgitation/insufficiency;
- Double tricuspid injury;
- Heart failure;
- Increase of RV Pd_2.
- Isolated Pulmonary Hypertension: the ECG is very sensitive in symptomatic patients with isolated pulmonary hypertension (**Bossone 2003**).

P wave of RAE in II and V1









Right Atrial Enlargement criteria (RAE)

I) Direct ECG criteria: Direct P wave criteria are very specific but their sensitivity is very low.

Voltage of P ≥2.5 mm in at least one of inferior leads: P pulmonale or P pulmonale parenchymal: tall and occasionally pointed P wave in inferior lea, Aspect in apex of P wave: “Goth P”, P wave height >1.5 mm in lead V_2. The criteria has 100% specificity preserved, P waves with “plus-minus” pattern in right precordial leads with initial plus component ≥1.5 mm, P wave deeply negative or positive in V_1 P wave of voltage ≥ at 1.5 mm in V_2 in association to R/S ratio >1, SÂP to the right of +80º (negative P wave in VL). In congenital heart diseases, SAP is not deviated to the right., P wave with increase in voltage and in duration in cases of extreme RAE, Macruz index lower than 1 = $^{\text{P duration}}/_{\text{PRs duration}}$ (**Macruz 1958**)

The QRS criteria with a QRS amplitude in V1 <4 mm + ratio V2/V1 > 5 are highly specific (>90%) with moderate SE (≈ 45 %)

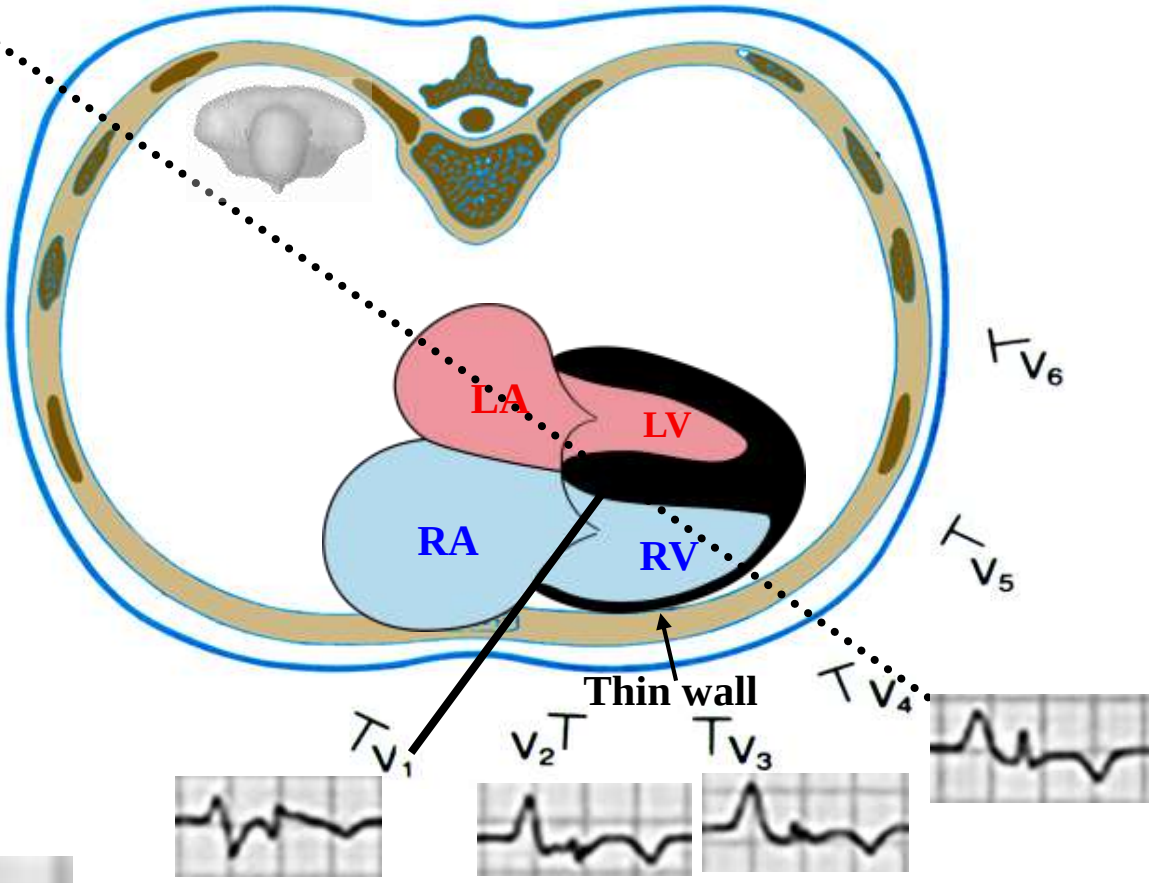
The combined P + QRS criteria with a P wave amplitude in V2 > 1.5 mm + SÂQRS > 90° + R/S ratio >1 in V1 in the absence of RBBB have 100% specificity and ≈ 50% sensitivity.

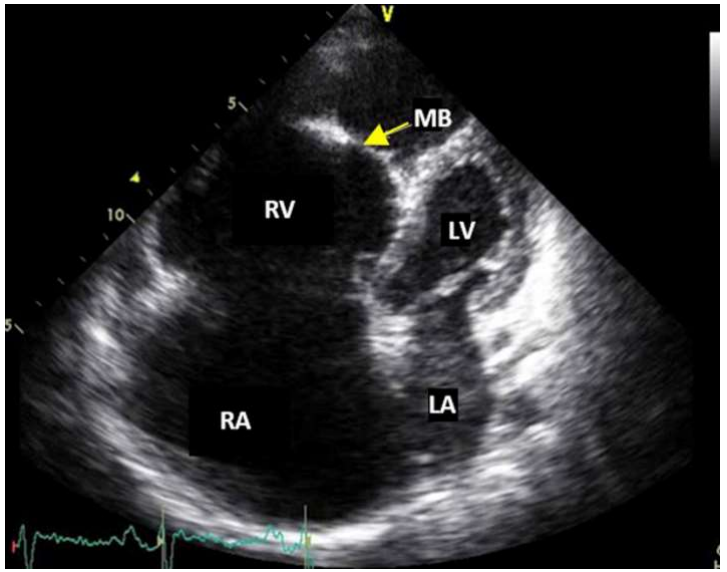
II) Indirect ECG criteria

$SÂQRS_F$: > 90º.The criteria has 100% specificity preserved, Sodi Pallares sign[1]: qR, QR or qRs in V_1 and V_2, Peñaloza and Tranchesi sign: QRS complexes of low voltage in V_1 contrasting with QRS complexes of normal voltage or increased in V_2., R/S ratio >1 in lead V_1 without RBBB. The criteria has 100% specificity preserved.

Hypothetical contrast-voltage among giant P waves and diminutive QRS complexes in the right precordial leads in the presence of Uhl's anomaly or parchment heart

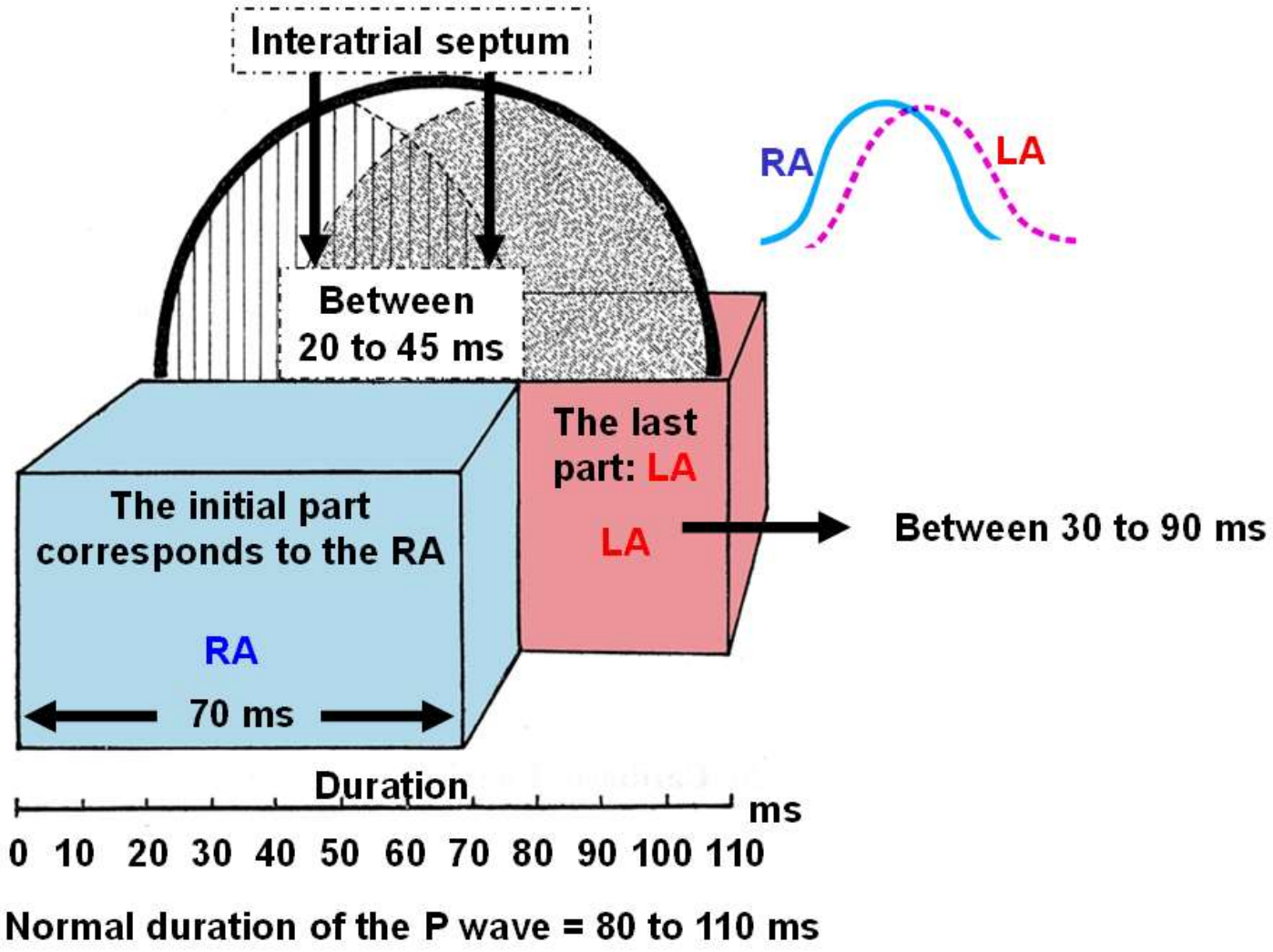
Big P wave **Small qrs**



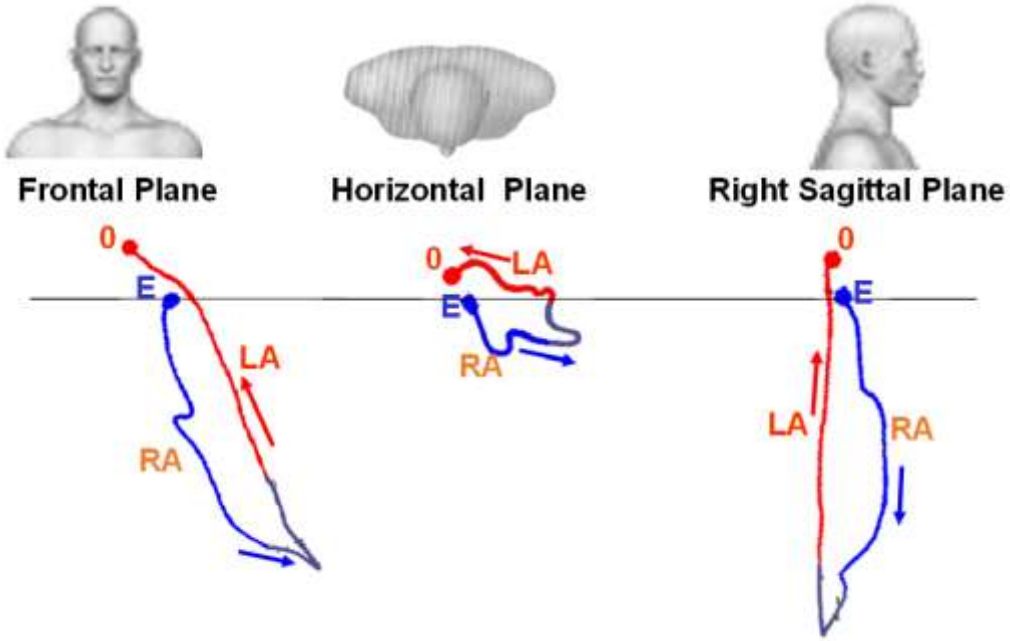


The extremely thin free wall of the RV is almost unable to produce electrical potentials on the ECG with minimal QRS amplitude in the right precordial leads which develops a parchment like appearance with associated diminution of function. The real cause of small QRS complexes from V1 to V3 is the **complete absence of the myocardium of the parietal wall of the RV.**

Normal sequential activation of biatrial chamber and interatrial septum

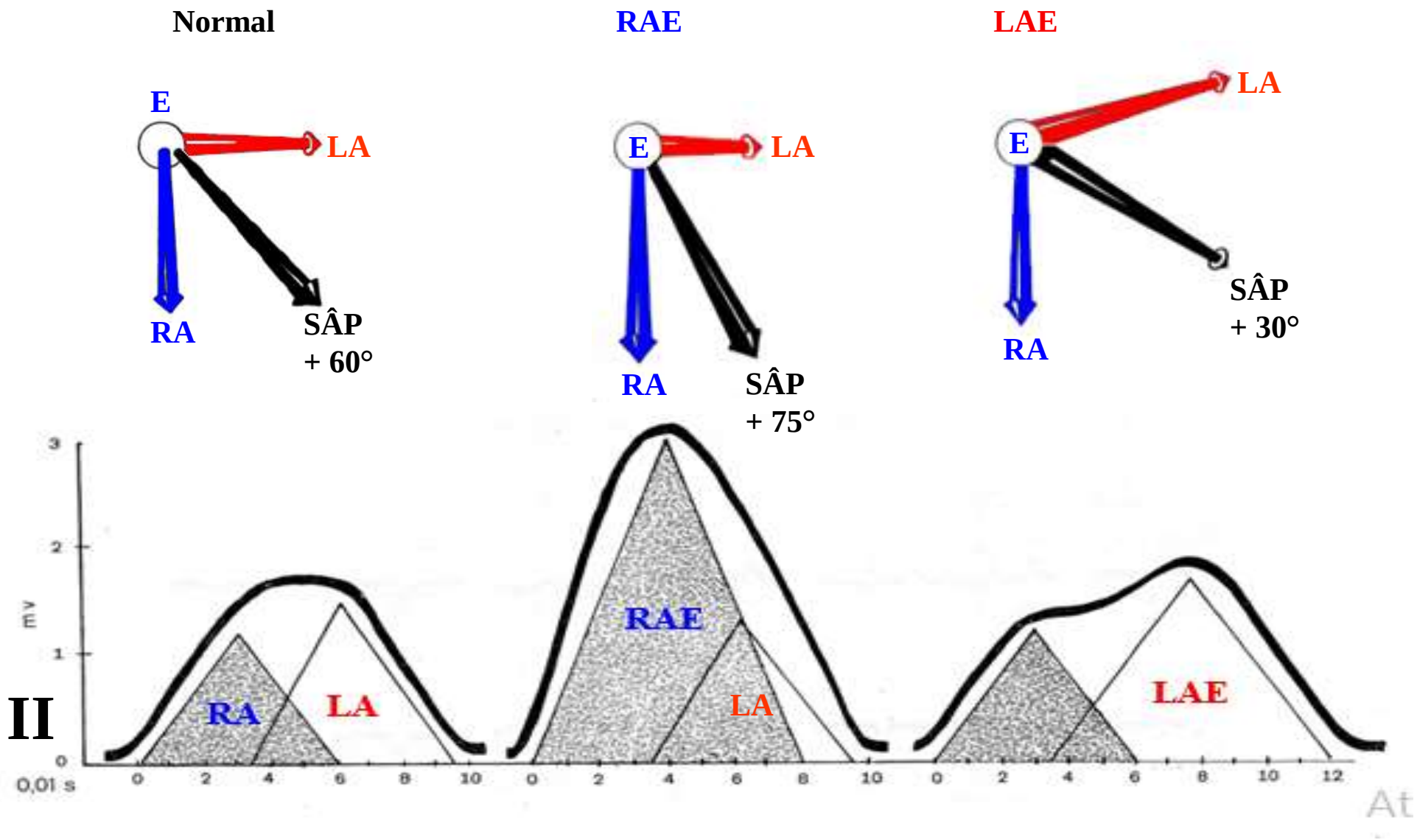


Normal P loop in the three planes



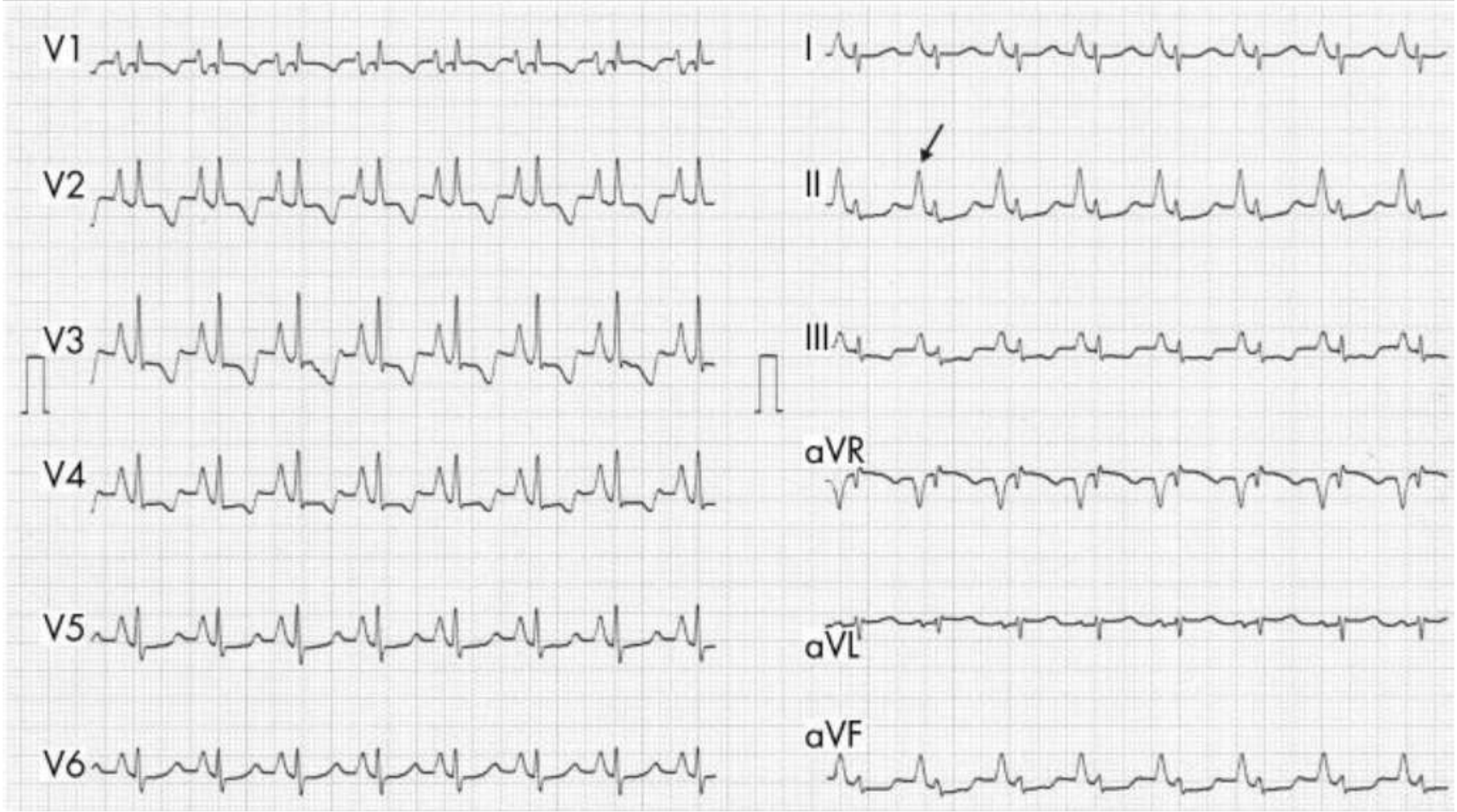
Normal sequence activation of biatrial chamber. The first 1/3 of the P wave corresponds to RA activation, the final 1/3 corresponds to LA activation; the middle 1/3 is a combination of the two chambers.

Profile of normal P wave in RAE and LAE

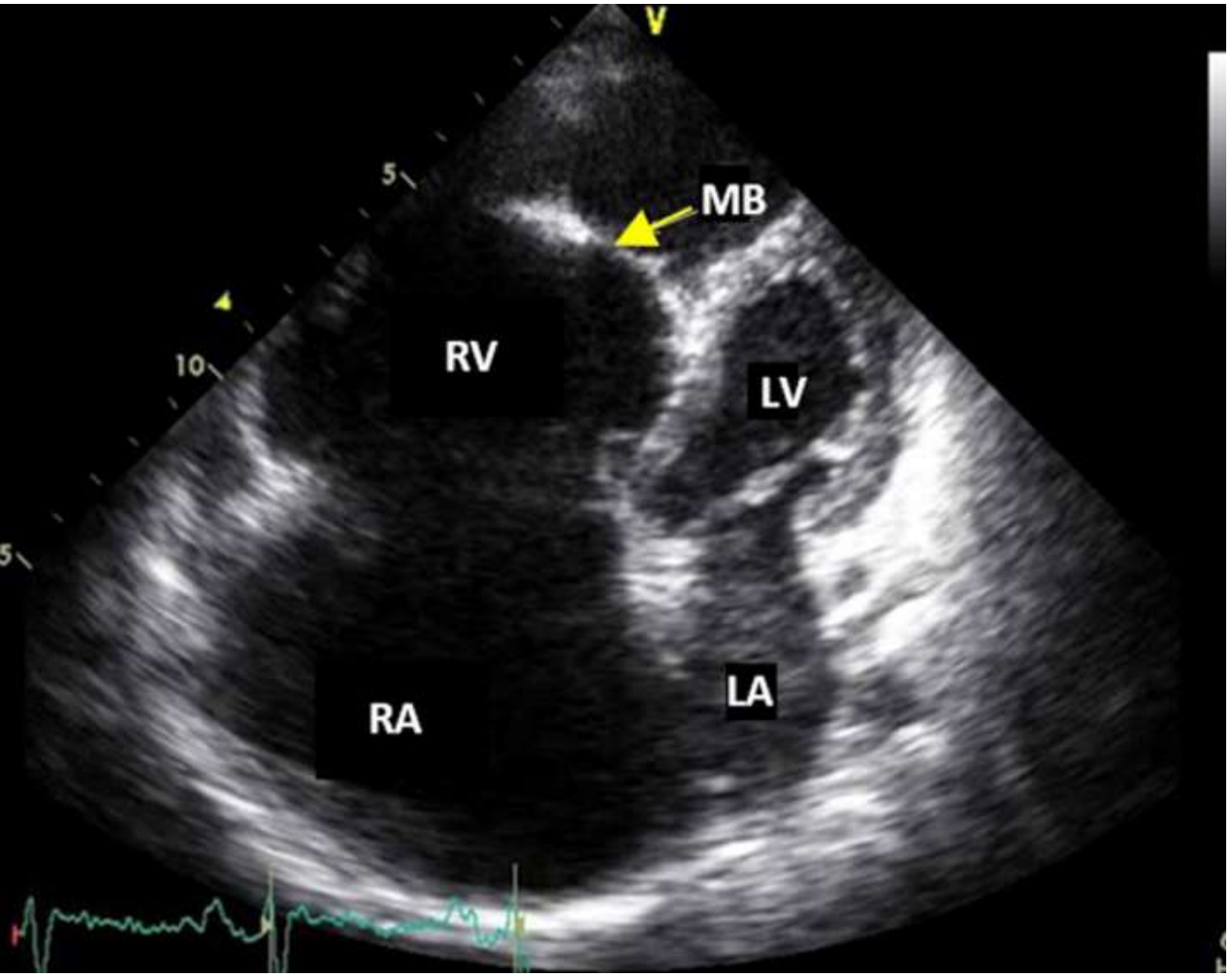


Causes of Himalayan P-waves or Alpine A Waves (modified from Peter P Vlismas 2019)

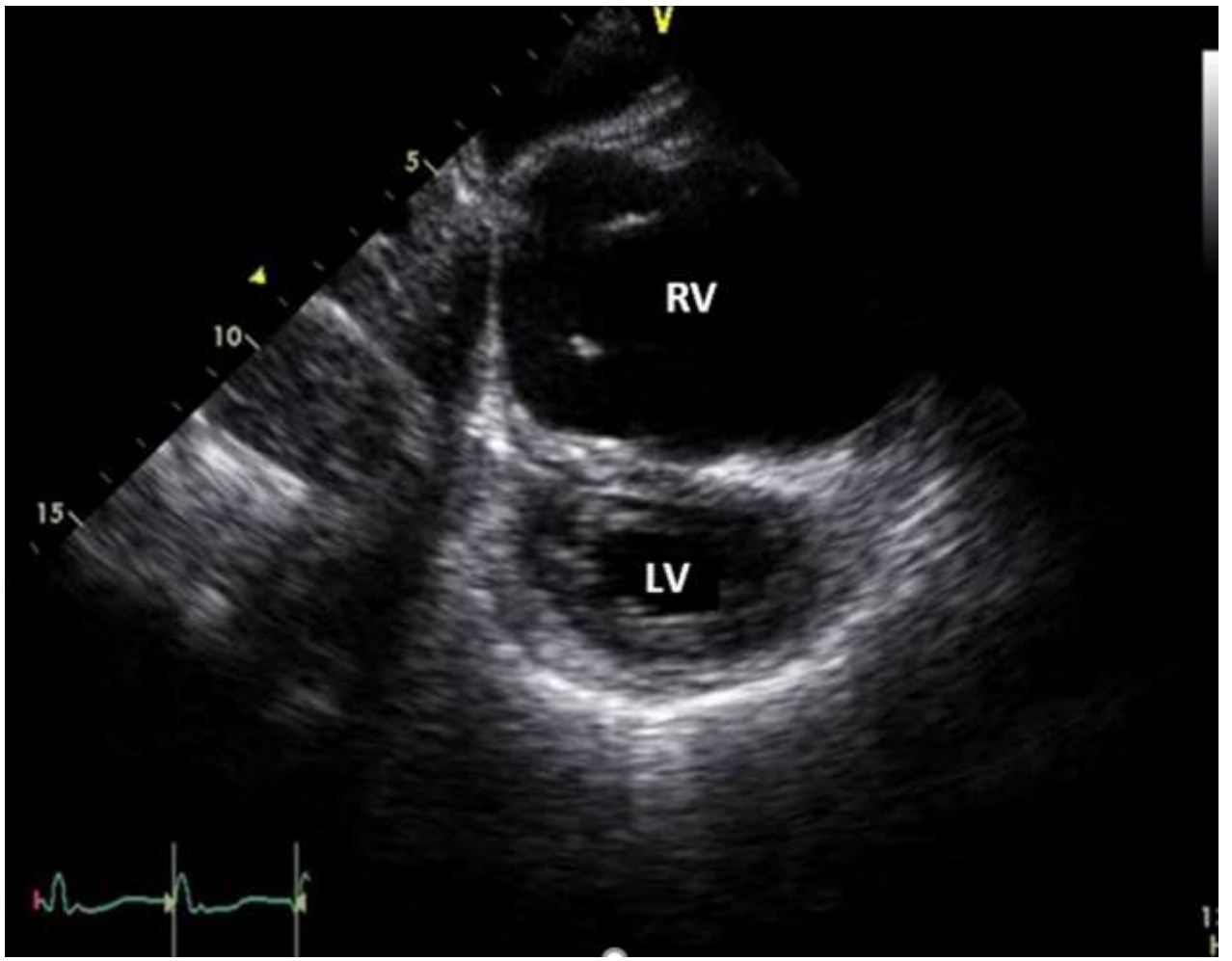
1. **Himalayan P-waves in a patient with tricuspid atresia (Reddy SC 2003) or** "P tricuspidale" or Gamboa P wave (**Gamboa 1966**). In this case Gamboa P wave is associated with diastolic, volumetric or eccentric left ventricular hypertrophy, left axis QRS deviation (LAD) in the frontal plane: LAFB pattern, counterclockwise rotation of QRS loop in the frontal plane: LAFB pattern, cyanotic baby (neonate or infant). It is very suggestive of tricuspid atresia diagnosis. Rosado-Buzzo et al from a cohort with 120 children with tricuspid atresia observed in their ECGs LAD in 94%, RAE in 58%, LAE in 47.5% and LVH in 96% (**Rosado-Buzzo 1987**).
2. **Restrictive cardiomyopathy. (Gupta MD, 2010.)**
3. **ischemic cardiomyopathy with triple vessel disease and four chambers were dilated with generalized hypokinesia of the LV (Sinha SK 2014).** In these cases the P waves are prominent in lead II, where it was even taller than the QRS.
4. **Uhl's anomaly or parchment heart (The present case)**
5. **Chronic obstructive pulmonary disease (COPD)** P-wave with rightward shift of P axis is the most discriminating P-wave change in evaluating the severity of COPD. **(Calatayud, J. B., Abad, J. M., Khoi, N. B., Stanbro, W. W., & Silver, H. M. (1970). *P-wave changes in chronic obstructive pulmonary disease. American Heart Journal, 79(4), 444–453.* doi:10.1016/0002-8703(70)90248-6).** Himalayan P-waves in COPD is a rare feature) Himalayan P-waves (P =5 mm) are often known to be classically associated with congenital heart diseases with right to left shunt like tricuspid atresia, Ebstein anomaly, combined tricuspid and pulmonic stenosis, etc, where they indicate a dilated RA and tend to be persistent. These type P waves are rarely seen in COPD and in this condition it may be due to structural RA changes or hypoxemia or combination of both. **(Kumar S 2013.)**
6. **Combined tricuspid and pulmonary stenosis (Davutoglu 2003).** See next slide.



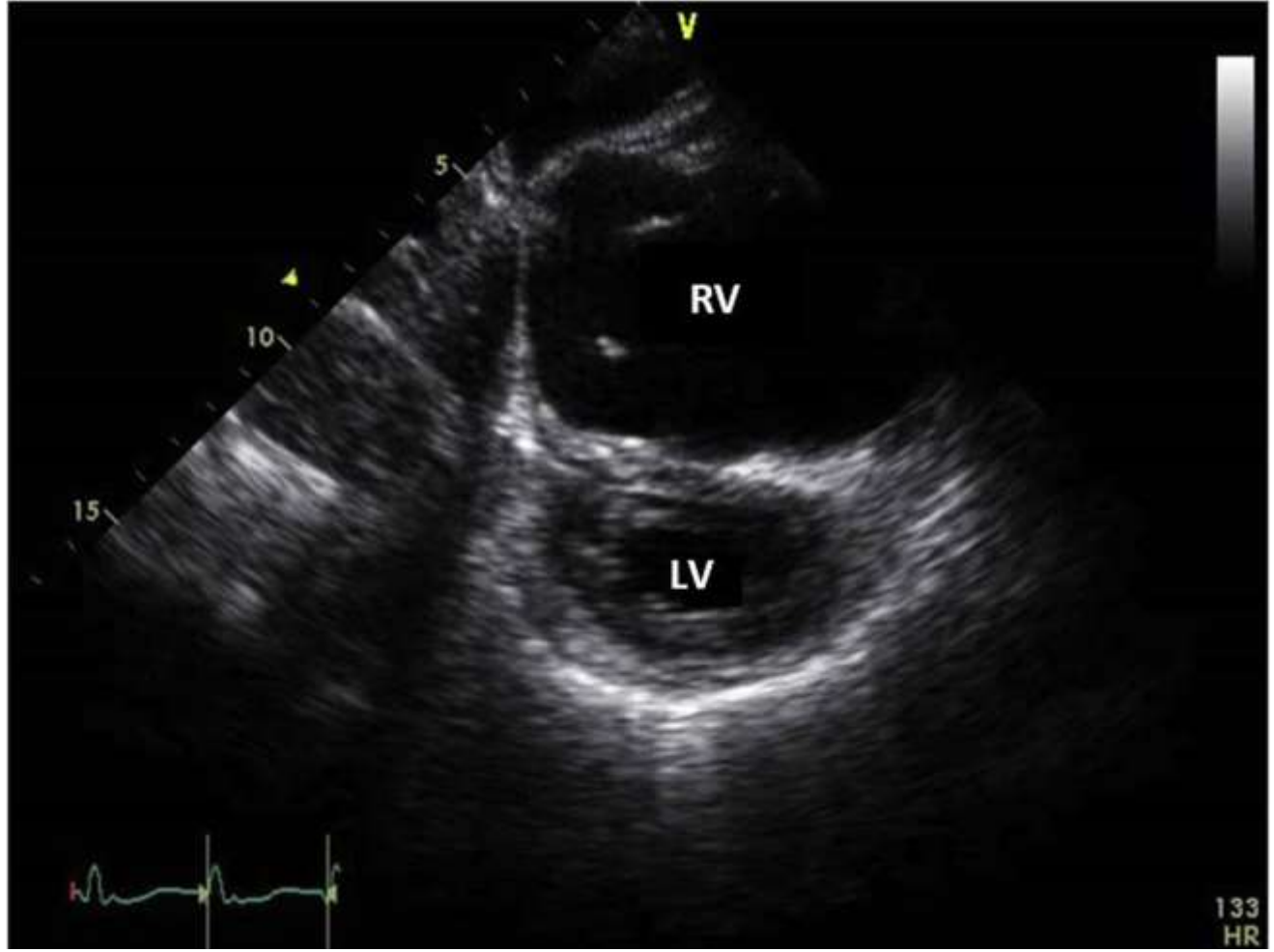
Clinical diagnosis: severe congenital combined tricuspid and pulmonic stenosis. 8-year-old female. Normal sinus rhythm, RAE, and RVH with right ventricular forces. The P waves are tall (> 5 mm) and peaked in lead II (black arrow). These types of P waves are called giant P waves or Himalayan P waves and are caused by reflected dilated RA resulting from pressure overloading.

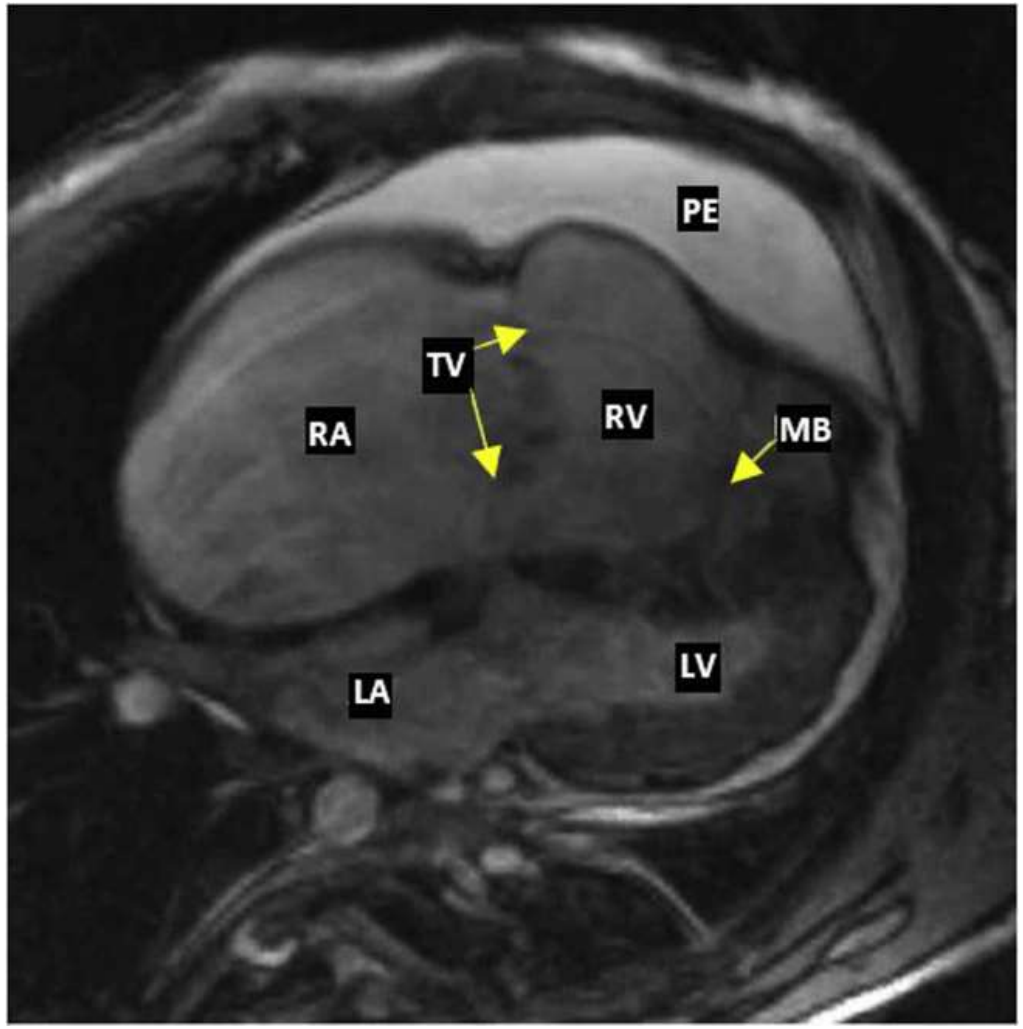


Transthoracic echocardiogram(TTE) , apical four-chamber view in diastole, showing marked right atrial and right ventricular dilatation. A moderator band (MB) and thin right ventricular walls were observed. *LA*, Left atrium; *LV*, left ventricle; *RA*, right atrium; *RV*, right ventricle.

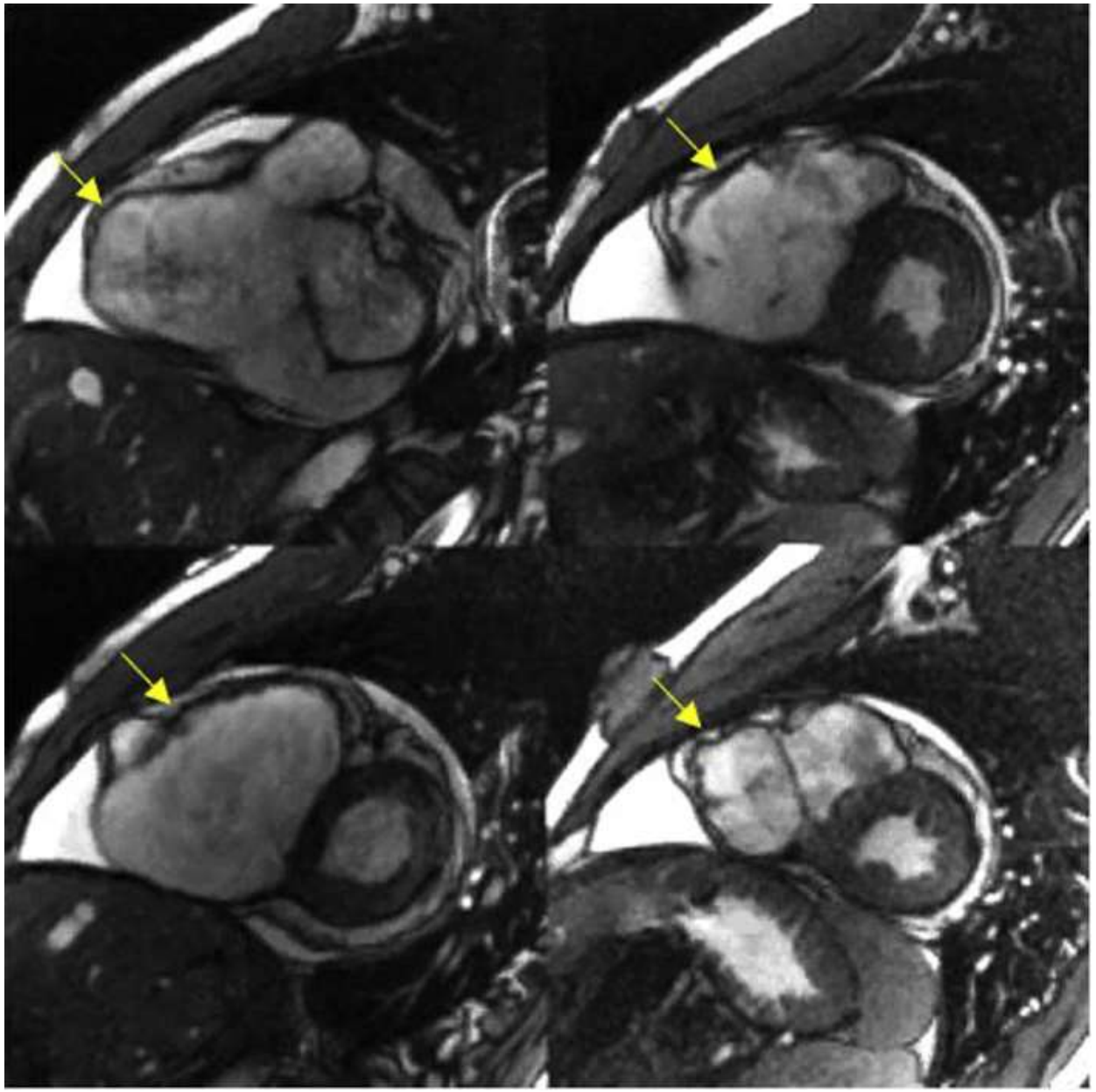


TTE, parasternal short-axis view in diastole, showing marked RV dilatation with thin walls and interventricular septum flattening. *LV*, Left ventricle; *RV*, right ventricle. TTE shows marked RV dilatation with thin walls (**1–2 mm** in almost all regions) and significant depression of its contractility. Right atrial enlargement is also observed, as well as hypertrophy with normal left chambers. The tricuspid valve has normal morphology and implantation.


V
5
10
15
RV
LV
133
HR



Cardiac magnetic resonance image(CMRI), in steady-state free precession, four-chamber sequence showing marked right chamber dilatation. The tricuspid valve (TV) has normal positioning. A moderator band (MB) and a medium-volume pericardial effusion (PE) are visualized. *LA*, Left atrium; *LV*, left ventricle; *RA*, right atrium; *RV*, right ventricle. Cardiac magnetic resonance imaging reveals an extremely thin-walled right ventricle with almost complete absence of free wall myocardium and with scarce apical trabeculations. There is no fibrofatty infiltration, with RV systolic dysfunction.



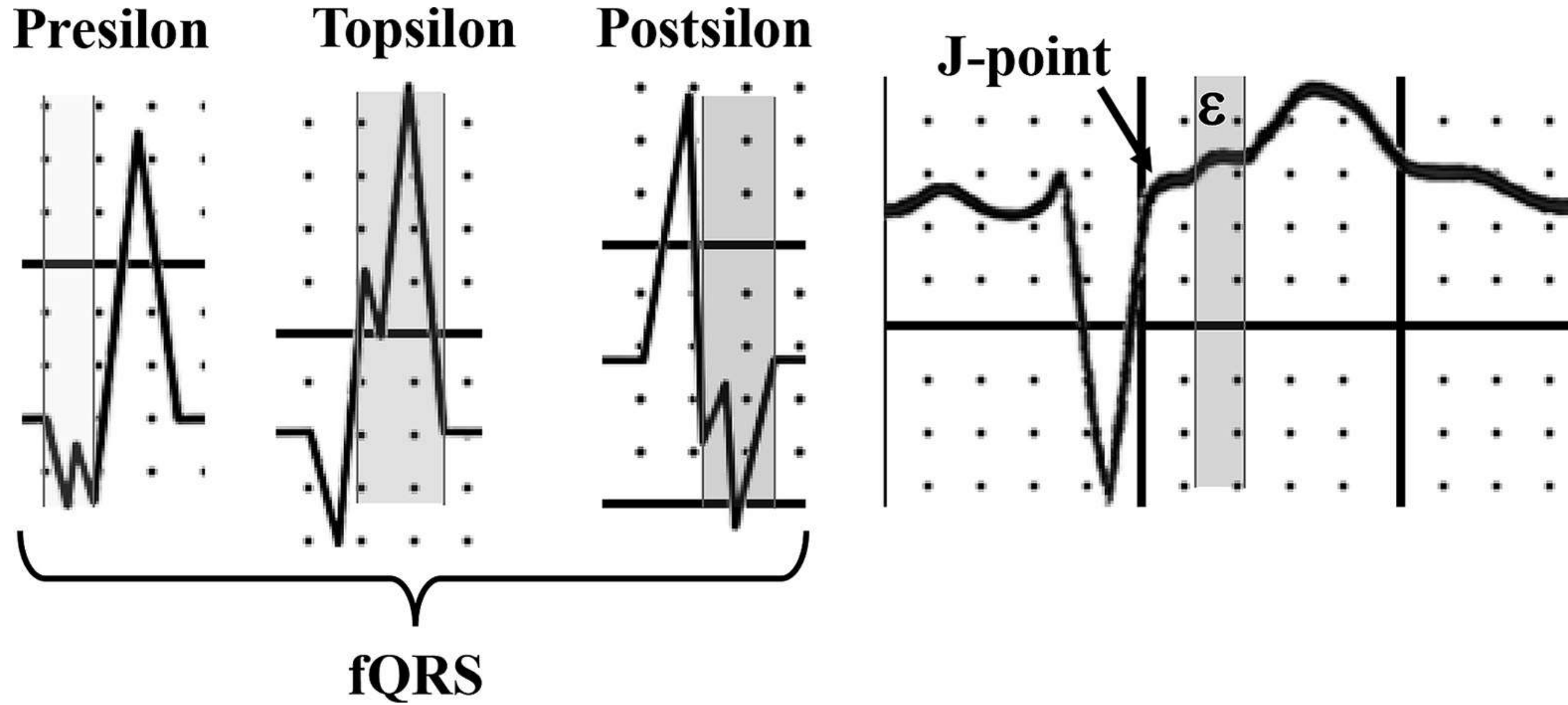
CMRI, in steady-state free precession, short-axis sequence showing normal left ventricular size, and the obvious thinning of the RV free wall (*arrows*).

In 1979, Fontaine *et al.* (**Fontaine 1979**) described ARVD, characterized by fibrofatty replacement of the RV myocardium.

Gerlis *et al.* concluded that many cases of ARVD were incorrectly classified as Uhl's anomaly, and these two entities had to be distinguished. (**Gerlis 2015**).

Table 1 shows the differential diagnosis between Uhl's anomaly or parchment heart and ARVC.

	Uhl's	ARVC
Family history	**No**	**Yes**
Sex ratio	**1:1**	**Polemic Conflict dates**
SCD	**No**	**Frequent in competitive Athletes**
Associated lesions	**1-4%**	**No.**
Age of presentation	Usually diagnosed in fetus, neonatal or infant life	Young adult. rarely manifest symptoms before the age of 20 years, and usually present with palpitations or else die suddenly.
	Complete absence of the myocardium of the parietal wall of the RV. No fatty tissue interposed between these layers.	**Fibro-fatty tissue replacement** of the parietal wall of the RV.
TVI	Posible	Right precordial leads TWI leads V1–V3 or beyond, aged>14 years of age, without complete RBBB
Epsilon waves	Prominent ε waves in all QRS complexes	



The figure shows the three possibilities of fragmented QRS in AC: at the beginning (presilon), in the middle (topsilon) and at the end (postsilon) of the QRS complex, and when the ε wave is located after the J-point and the beginning of the ST-segment. Although the ε wave is a depolarization abnormality (late potential), it is recorded at the beginning of repolarization.

Uhl's anomaly is usually diagnosed in neonatal or infant life and present with congestive heart failure. It is a very rare congenital abnormality characterized by the almost complete absence of the myocardium in the RV which develops a parchment like appearance with associated diminution of function. It was first described in 1952 by Henry Uhl after performing an autopsy on an 8-month-old infant (**Uhl 1952**), and it is thought that fewer than 50 cases have since been described. During embryonic development the human heart advances through phased embryological processes. It is thought that loss of the RV myocardium must only occur after complete cardiac development. Apoptosis is a routine component of postnatal morphogenesis of the human heart and it has been speculated that unrestrained RV myocardial apoptosis may be responsible (**James 1994**). With advancing imaging capabilities, now exemplified by cardiac magnetic resonance (CMR) imaging, the diagnosis can be made more readily providing an opportunity to intervene surgically. Patients often present in infancy and rarely survive to adulthood without intervention. The typical imaging findings consist of a thinned, akinetic RV wall in association with a paucity of trabeculation with a dilated RV cavity (**Greer 2000**).

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