

CLINICAL CASE:  
**SEVERE IDIOPATHIC PULMONARY  
HYPERTENSION IN CHILDREN AT ALTITUDE**

**The importance of early detection**

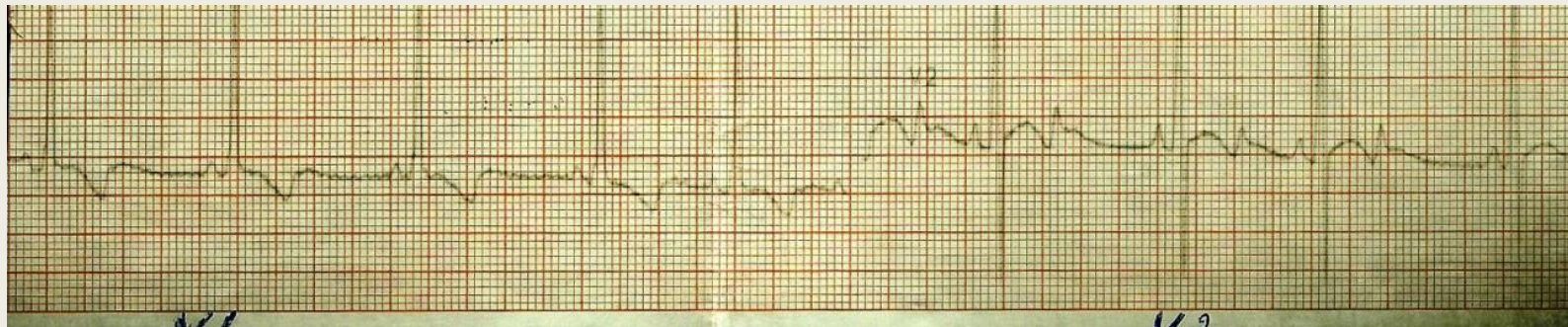
---

**GABRIEL F. DIAZ G.**

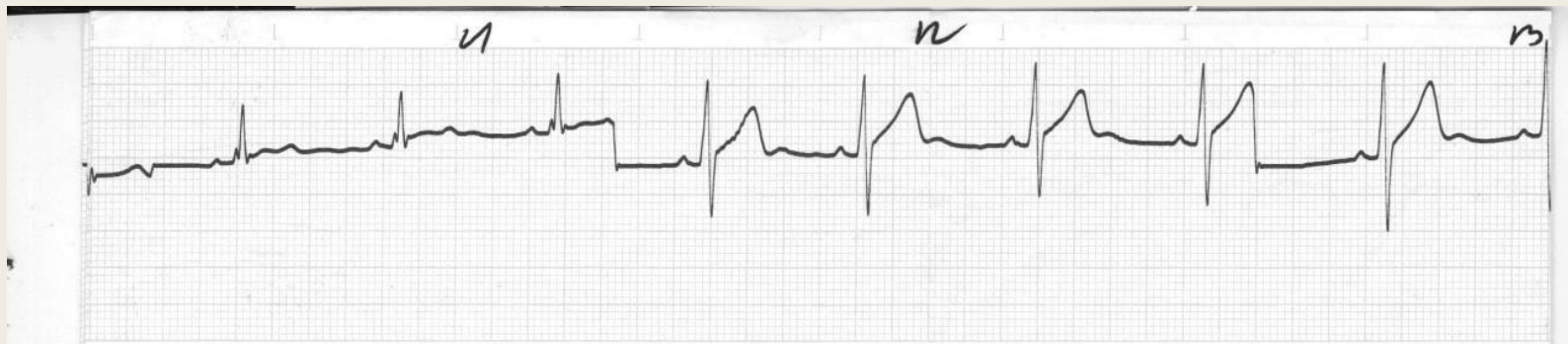
- TITULAR PROFESSOR, DEPARTMENT OF PEDIATRICS  
UNIVERSIDAD NACIONAL DE COLOMBIA
- PEDIATRIC CARDIOLOGIST, FUNDACIÓN SANTAFE DE BOGOTA

- 21 MONTHS OLD PATIENT. DUE TO A MURMR, A THORAX X-RAY WAS SOLICITED
- **THORAX X-RAY:** IMPORTANT CARDIOMEGALY (RV, RA, AND PT ENLARGEMENT)
- **CARDIAC EVALUATION:** MILD CIANOSIS, NORMAL PULSES, NO HEPATOMEGALY, CARDIAC HYPERACTIVITY, LOUD SECOND HEART SOUND, PROTOSYSTOLIC CLICK AND SYSTOLIC MUMUR II/VI IN STERNAL BORDER
- **ECHOCARDIOGRAM:** SEVERE PH (153 MM PH), WITH DILATION OF RV, RA AND PULMONARY TRUNK
- **EKG:** IMPORTANT HYPERTROPHY OF THE RV
- **DIAGNOSIS:** SEVERE IPH
- PROLONGED HYPEROXIA TEST (PHT) WAS (+). PSP DIMINISHED UNTIL 94 MM PH

- **CATHETERISM:** SEVERE IPH WITH VRT (-). ANY POSSIBILITY OF TREATMENT WAS DISCARDED
- DUE TO +PHT, LIVING AT LOW ALTITUD WITH SILDENAFIL WAS RECOMMENDED
- THE PATIENT HAD 3 SYNCOPES IN 15 DAYS. BETTER PROGRESSIVELY AT LOW ALTITUDE. O2 WAS WITHDRAWED SLOWLY. ONE MONTH AFTER LIVING AT LOW ALTITUDE, PATIENT STOPPED NEEDING O2. CONTINUED LIVING AT LOW ALTITUDE UNTIL TODAY (18 YEARS OLD)
- CONTINUED WITH SILDNAFIL. HE RECEIVED SILDENAFIL PLUS BOSENTAN FOR 6 MOTNHS, IT WAS WITHDRAWED PROGRESIVELY
- HE WAS ABLE TO HAVE ALL THE SAME PHYSICAL ACTIVITIES AS HIS CLASSMATES AT SCHOOL
- HE IS NOW 18 YEARS OLD AND GOING TO UNIVERSITY
- IN A PILOT STUDY LOOKING FOR BMPR2 MUTATIONS (19 PATIENTS), HE WAS THE **ONLY PATIENT WITH A MUTATION:** A VALINE FOR AN ALANINE

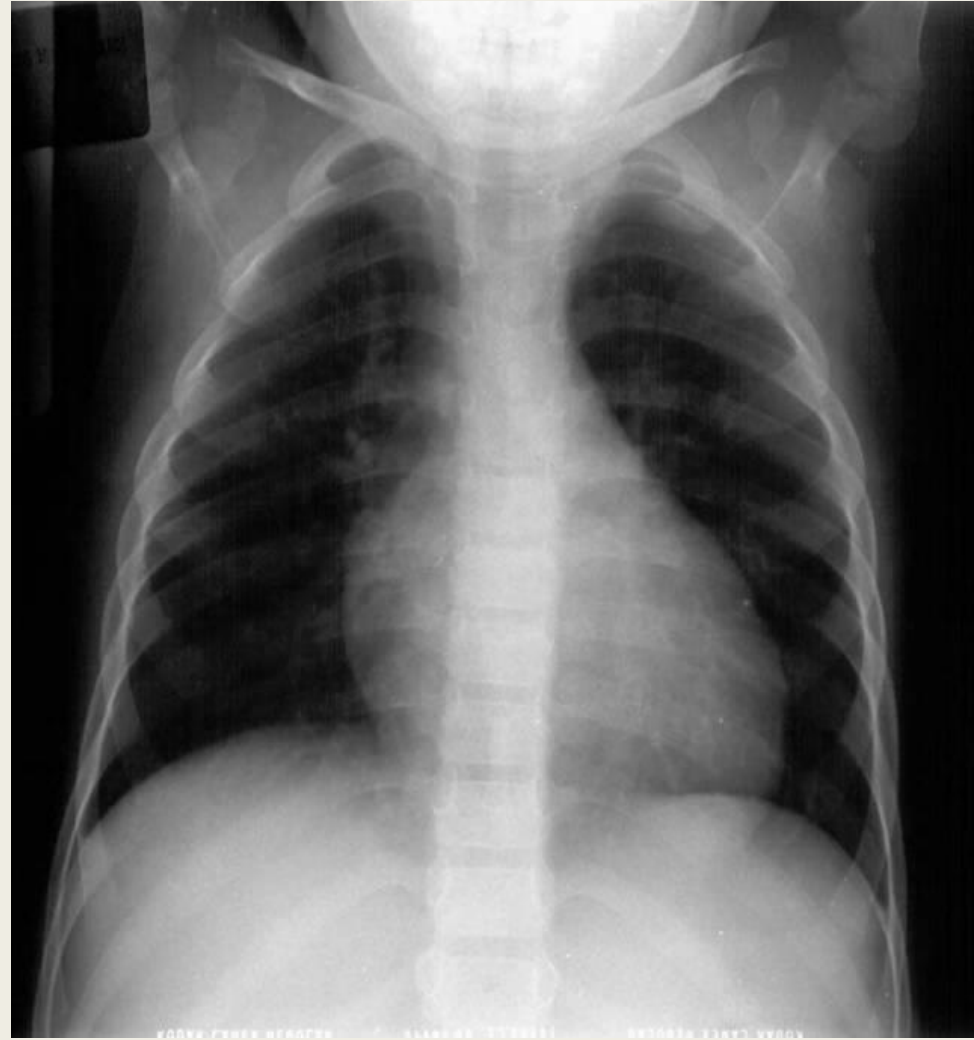


**21 MONTHS OLD**



**17 YEARS OLD**

- $\frac{1}{2}$  VOLTAGE (IN BOTH EKG)
- THE SUPERIOR PANEL IS THE INITIAL EKG WHEN THE PATIENT WAS 21 MONTHS OLD. THE R IN V1 IS 44 MM
- THE INFERIOR PANEL IS THE ACTUAL EKG AFTER 17 YEARS OF FOLLOW UP. THE R IN V1 IS 14 MM



SEVERE IDIOPATHIC PULMONARY HYPERTENSION IN CHILDREN AT ALTITUDE  
The importance of early detection

## **INITIAL CATHETERISM**

BASAL: PP: 84 /20; 48 PR: 12 WU

**VRT (HYPEROXIA, NITROPRUSSIDE AND ISOPROTERENOL) : NEGATIVE**

## **CATHETERISM (ONE MONTH AGO):**

BASAL: PP: 54/16, 32; PR: 3.62 WU; SR: 18.87 WU

OXYGEN: PP: 45/25, 23 PR: 0.67 WU; SR: 11.67 WU

ILHOPROST: PP: 49/14; 25 PR: 1.18 WU; SR: 13,58 WU

**BNP: 60 Pg/ml**

GILBERTO ESTRADA M.D.  
IGNACIO CALDERON M.D.  
PABLO CASTRO M.D.  
GERMAN GÓMEZ M.D.  
JORGE MOR M.D.  
EDGAR HURTADO M.D.

# FUNDACION CLINICA SHAI0

DEPARTAMENTO DE HEMODINAMIA  
CARDIOLOGIA INTERVENCIONISTA



848509

0

66.002

2

15-May-02

Compensar E.p.s.

PACIENTE BOHORQUEZ MOYA ANDRES FELIPE

INDICACION:

CONTROL MEDICO

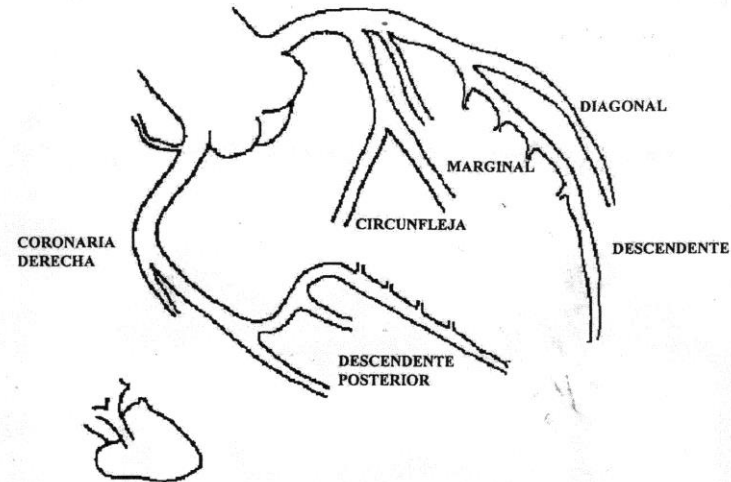
Historia

Estudio

Edad

Fecha

Entidad



F.E. 60%

### Comentario

PROCEDIMIENTO: CATETERISMO DERECHO E IZQUIERDO, VENTRICULOGRAMA AORTOGRAMA, HIPEROXIA, ADMINISTRACION DE NITROPRUSIATO, CAPILAR PULMONAR. VENTRICULO IZQUIERDO DE TAMAÑO NORMAL, CONTRACTILIDAD NORMAL DE TODAS SUS NPAREDES, VALVULA MITRAL COMPETRENTE. AORTOGRAMA: VALVULA AORTICA TRIVALVA CON APERTURA Y CIERRE NORMALES, ORIGEN NORMAL DE LAS ARTERIAS CORONARIAS, SE DESCARTA COARTACION Y DUCTUS. ANGIOGRAFIA PULMONAR NORMAL, ANGIO PULMONAR GRADO III. DRENAJE VENOSO PULMONAR NORMAL.

Jorge Tibbo Mor Delle  
19 264 925 Bogotá  
Clínica Shaio

Drs: G. Estrada, L. Calderon, P. Castro, G. Gomez, J. Mor, E. Hurtado

PARÁMETROS	BASALES	FINALES	1 MINUTOS	5 MINUTOS	10 MINUTOS
FIO <sub>2</sub> (Litros/minuto)	21%	21%	21%	21%	21%
Saturación de oxígeno (%)	98%	94%	96%	97%	97%
Frecuencia Cardíaca (lpm)	72	86	94	89	90
Frecuencia Respiratoria (rpm)	17	23	21	15	20
Tensión Arterial (mmHg)	142/87	150/75	150/72	140/83	140/80
Disnea/Fatiga (Borg)	1	2	1	1	1

Distancia recorrida (Metros): 480 mts	Número de Detenciones: 0	Tiempo Total: 6 MINUTOS
VO <sub>2</sub> Máximo: 26		
Ortiz/Hoyos/Valencia: 685 m	70% Predicho:	METS: 7.42
Doble producto (RRR): 27300		Índice de gasto energético (EEI): 0.41

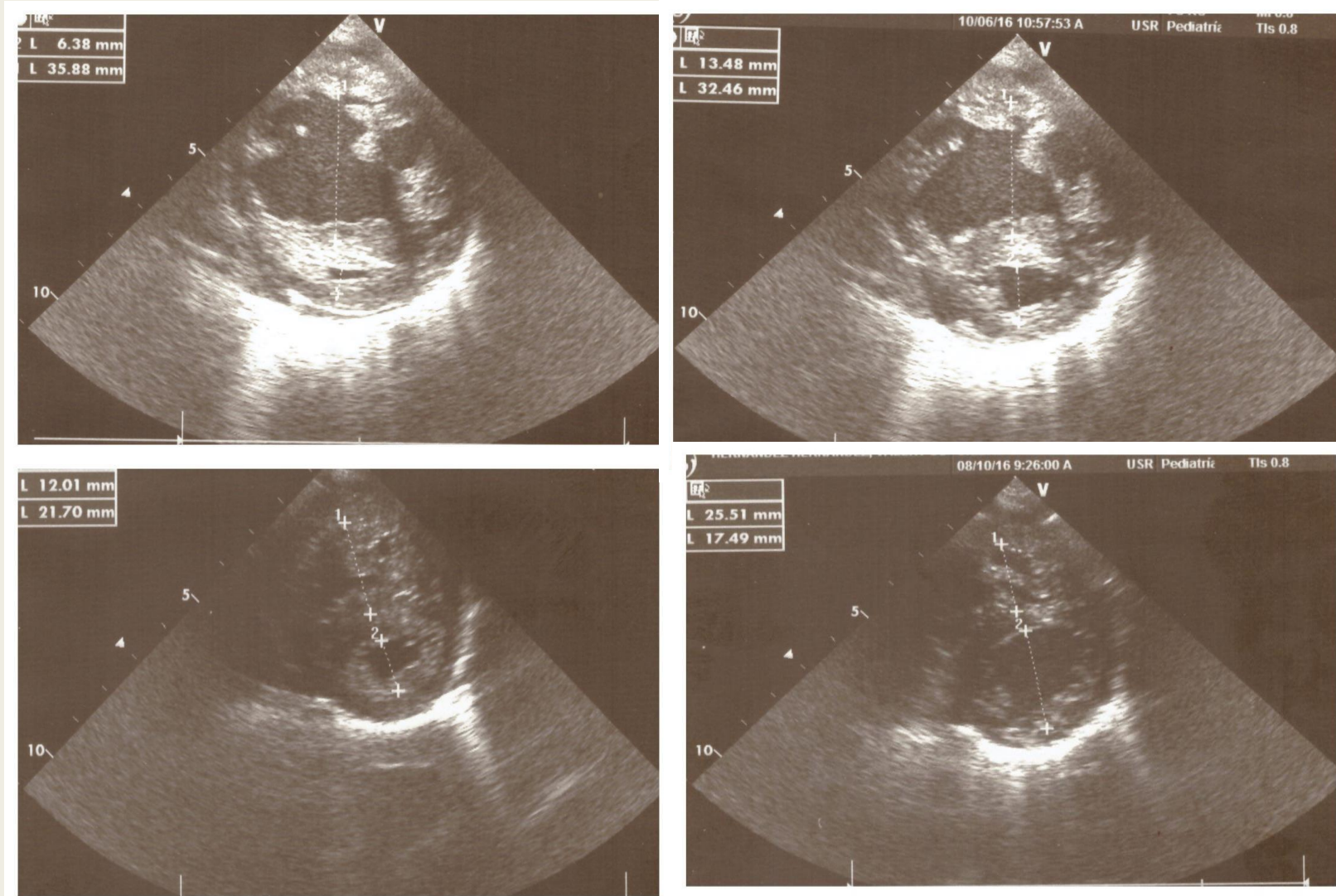
Escala de Borg modificada (0-10) mayor puntaje peores síntomas.

Lin: límite inferior de normalidad



## CONCLUSIONS:

- In our experience, PH in children at early age is frequent at altitude
- May debut as severe PH
- Hyperreactivity of pulmonary vasculature is significant at altitude. For its valoration we use the Prologed Hyperoxia Test
- **Early detection is very important** in order to avoid the remodeling of pulmonary vasculature
- Part of treatment is to live at low altitude



SEVERE IDIOPATHIC PULMONARY HYPERTENSION IN CHILDREN AT ALTITUDE  
The importance of early detection