

PREGNANCY IN A PHACE SYNDROME AND CONGENITAL HEART DISEASE PATIENT

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**The 4th International Congress
on Cardiac Problems in Pregnancy (CPP2016)**
27 Feb – 1 March 2016
LAS VEGAS, Nevada, USA



Our experience

- “Pregnancy and Heart disease” multidisciplinary team since 2009
- 102 patients with congenital heart disease



This is the story of Cristina...

- 34 yo woman
- PHACE syndrome
- Operated congenital heart disease
- Arterial hypertension
- Therapy: amlodipine 10 mg

We met Cristina at the 15th week of gestational age

PHACE Syndrome

Pediatric Neuroradiology

Neuroradiology

December 1978, Volume 16, Issue 1, pp 82-84

First online:

Vascular and nonvascular intracranial malformations associated with external capillary hemangiomas

I. Pascual-Castroviejo

Ped Neurorad, 1978

PHACE Syndrome

Article | March 1996

PHACE Syndrome

The Association of Posterior Fossa Brain Malformations, Hemangiomas, Arterial Anomalies, Coarctation of the Aorta and Cardiac Defects, and Eye Abnormalities

Ilona J. Frieden, MD; Vail Reese, MD; Debra Cohen, MD

Arch Dermatol. 1996;132(3):307-311. doi:10.1001/archderm.1996.03890270083012.

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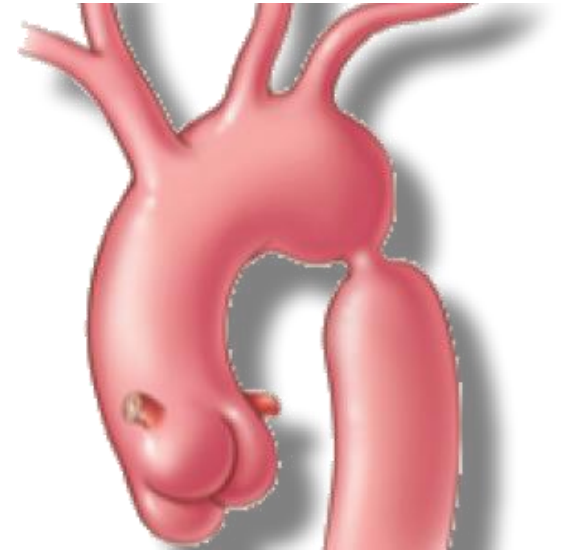
Infants diagnosed with PHACE syndrome may have only one or two of these abnormalities, in addition to the hemangioma.

PHACE Syndrome



Aortic Arch anomalies

Around 40% of children with PHACE syndrome have aortic arch anomalies, including coarctation and aortic interruption.



Chest. 1982 Aug;82(2):186-7.

Coarctation of the aorta with congenital hemangioma of the face and neck and aneurysm or dilatation of a subclavian or innominate artery. A new syndrome?

Schneeweiss A, Blieden LC, Shem-Tov A, Motro M, Feigel A, Neufeld HN.

Cristina

- She was born in 1981.
- Extended emangiomas of the cervicofacial region and of the larynx treated by tracheostomy
- Corticosteroid therapy was started
- 1982→ gastrostomy for 2 months
- A big cutaneous angioma of the back was treated by surgery
- Congenital hypothyroidism
- Echocardiography (1982): negative

Poor prognosis

Cristina

- Several hospitalizations
- 1984: partial regression of hemangiomas → tracheal cannula removed
- 1988: Diagnosis of hypertension

Echo: aortic coarctation

1988, Surgery

CENTRE HOSPITALIER UNIVERSITAIRE DE LA TIMONE

HOPITAL DES ENFANTS

1, Boulevard Jean Moulin - 13385 MARSEILLE CEDEX 5

Les examens qu'elle a subi dans notre hopital ont montré qu'elle présentait une anomalie très particulière, c'est à dire un double arc aortique avec hypoplasie de l'arc aortique droit et postérieur et coarctation aortique sur l'arc aortique gauche.

- Rare anomaly
- Double aortic arch
- Left dominant arch interrupted
- Right Hypoplastic arch

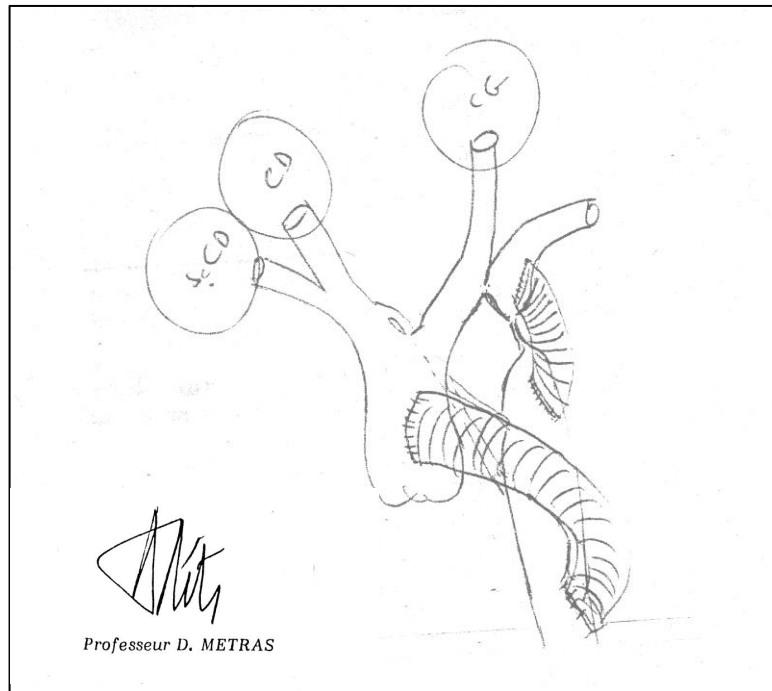
- Dacron Bypass of 12 mm between left arch and descending aorta

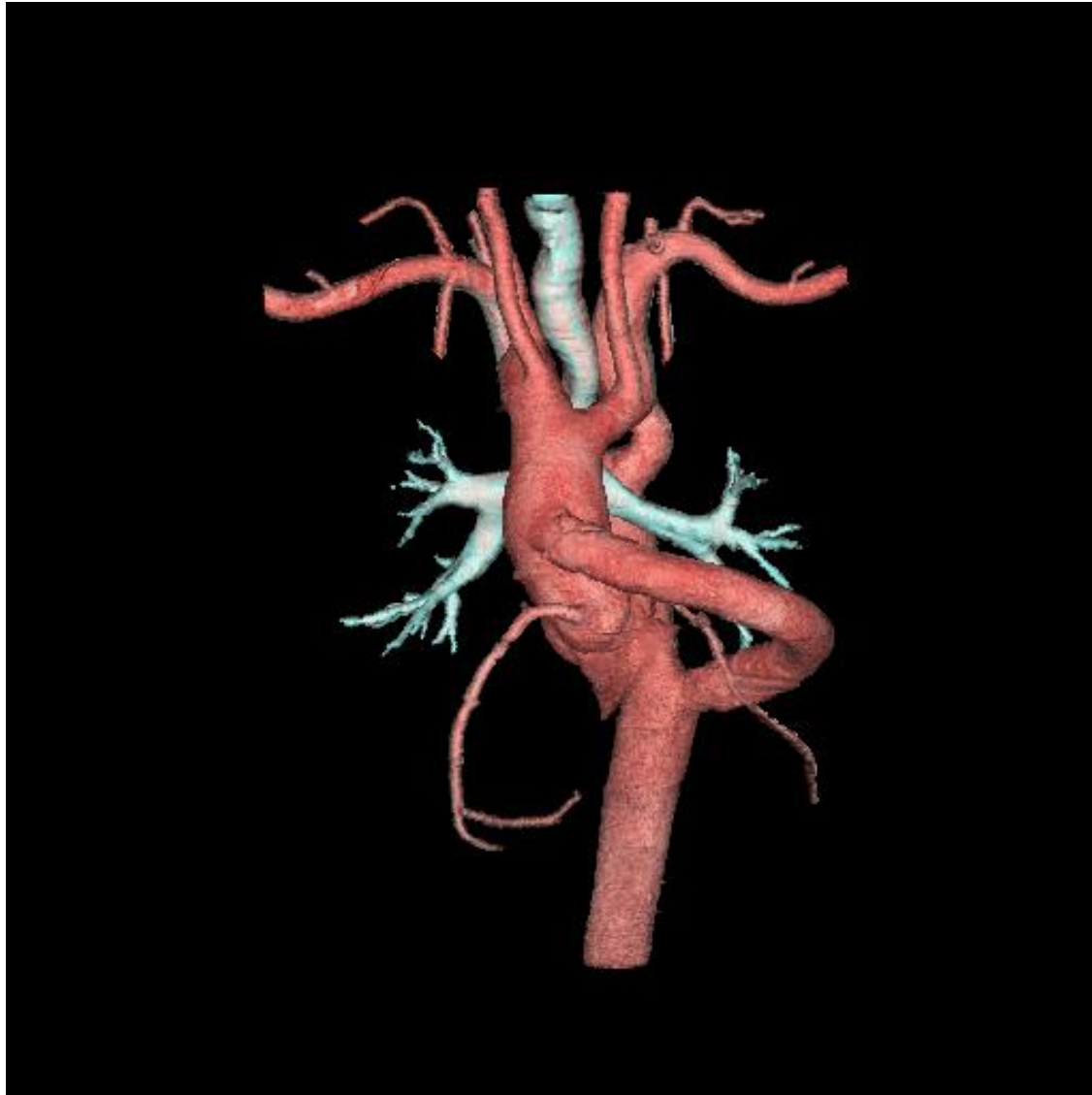


Professeur D. METRAS

1992, second aorto-aortic bypass

- Hypertension
- Dacron 14 mm bypass between ascending aorta and thoracic aorta





Thanks to D.Marini and F. Ferroni

Pregnancy

LMP:14/9/2014 DD: 21/06/2015

- Asymptomatic
- NYHA class I WHO II-III
- PA 135/90 mmHg. No pressure gradient between right arm and legs. Pressure gradient was seen between the two arms due to arch anatomy



Cardiologist



OBGyn



Anesthesiologist



Neurologist



Internist



Genetist



Pregnancy – Heart disease

- First trimester echocardiography reported a left ventricle mild hypertrophy with normal systolic function. Bypasses were difficult to be examined by ultrasounds
- Normal follow up resulted also during the second trimester

Arterial Hypertension

Pregnancy – Third trimester

- Uncontrolled arterial pressure, needing a hospital admission.
- A combined therapy (nifedipine, metoprolol, alphametildopa) was started and reached satisfactory values
- Holter blood pressure was normal at rest, with peaks up to 190/105 mmHg during emotional stress



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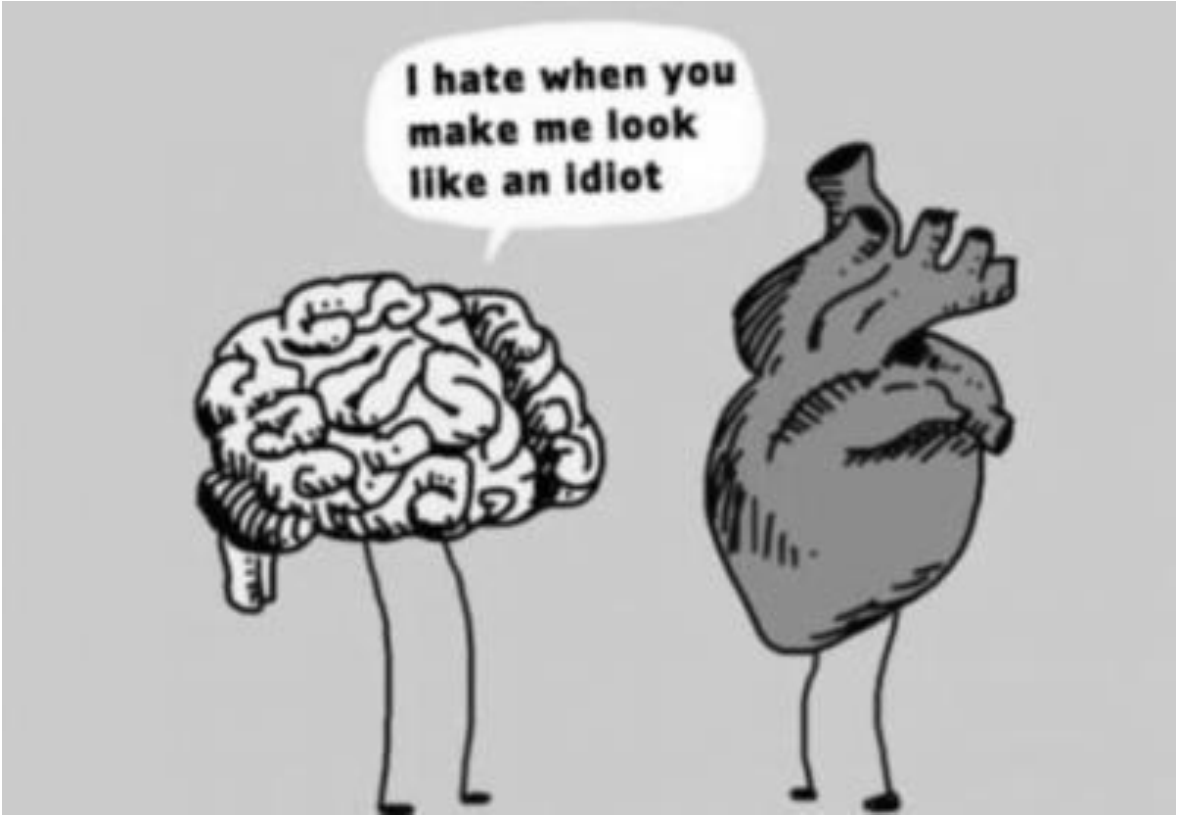
European Heart Journal (2005) 26, 2173–2178
doi:10.1093/eurheartj/ehi338

Clinical research

Outcome of pregnancy in patients after repair of aortic coarctation

22% were complicated by a hypertensive disorder of pregnancy.

Vriend, Eur Heart journal



Pregnancy

Increased risk of cerebrovascular anomalies

Cerebral MRI was always refused by the patient

115 PHACES cases, 89 (77.4%) had congenital and/or progressive cerebral vascular anomalies:

- dysplasia
- aberrant origin or course,
- hypoplasia, and absence or agenesis.
- Arterial occlusions and stenoses 20.9% and 18.3%
- embryonic arteries 17.4%
- saccular aneurysms 15 %

Delivery

- Due to hypertensive peaks and the unknown cerebrovascular anatomy an elective caesarean section was scheduled
- The baby was born at the 38 week of gestational age
- F, 2860 g APGAR 9/9. No malformations
- After delivery Cristina was asymptomatic
- Antihypertensive therapy was arranged



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Thank you!!!

Comoglio FM, MD

Pagano A, MD

Macchi C, MD

Donvito V, MD

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Agnoletti G, MD

Prof. Todros T, MD