PREGNANCY IN A PHACE SYNDROME AND CONGENITAL HEART DISEASE PATIENT

Bordese Roberto, MD
Pediatric Cardiology department
Città della Salute, Turin - Italy
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

Vascular and nonvascular intracranial malformations associated with external capillary hemangiomas

I. Pascual-Castroviejo

Ped Neurorad, 1978
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.


_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

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

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

Outcome of pregnancy in patients after repair of aortic coarctation

22% were complicated by a hypertensive disorder of pregnancy.

Vriend, Eur Heart journal
I hate when you make me look like an idiot.
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 %

Heyer, Stroke 2007
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
Thank you!!!