CASE REPORTS

SUCCESSFUL PREGNANCY IN A PATIENT WITH EISENMENGER’S SYNDROME (ATRIAL SEPTAL DEFECT WITH SHUNT REVERSAL)

S. H. P. NANAYAKKARA, F.R.C.S. (Eng.), M.R.C.O.G.
Visiting Obstetrician and Gynaecologist, Castle Street Hospital for Women, Colombo.

E. V. PIERIS, M.D., M.R.C.P., M.R.C.P. (Edin.)
Visiting Physician, General Hospital, Colombo, Ceylon.

Maude Abbott (1936) stated that the term Eisenmenger complex was used by her, for want of a better, to designate an unusual combination of VSD with dextroposition of the aorta without pulmonary stenosis or hypoplasia—first diagnosed during life by Von Schrötter and reported by Eisenmenger (1897).

Wood (1950) suggested that pulmonary hypertension with reversed aorto-pulmonary, interventricular or interatrial shunt might be called acquired Eisenmenger's syndrome because it is indistinguishable from Eisenmenger's complex. In his Croonian lectures on the syndrome (1958) he elaborated on this theme and used the term Eisenmenger syndrome for the clinical condition as a whole, the term Eisenmenger complex being reserved for the variety where shunt was through a ventricular septal defect, which was described first by Eisenmenger (1897).

The nomenclature used by Wood (1958) does not appear to have gained wide acceptance. Taussig (1960) states that it is advisable to regard the Eisenmenger complex as a syndrome defined as a heart with a large VSD, increased pulmonary blood flow and high pulmonary pressure.

Burwell and Metcalfe (1958) describe pregnancy in a case of cyanotic Eisenmenger's complex. (Shunt through a VSD.)

Carter (1948) mentions pregnancy and abortion in a case of Eisenmenger's syndrome—which he defines as a ventricular septal defect and dextroposed over riding aorta.

Case Report

Primigravida, aged 24 years, first seen on 14.6.61, when sixteen weeks pregnant, complaining of dyspnea for ten years, angina and syncopal attacks on exertion. She had also three hemorrhages in 1960. Expected date of delivery 19.11.61.

Past History—She was diagnosed at the General Hospital, Colombo as having an ASD in July, 1959. She entered Hammersmith Hospital on 18.1.60, for investigation. She was diagnosed as Eisenmenger complex (ASD with reversed shunt).

On examination—No dyspnea at rest; central cyanosis with clubbing of fingers and toes, pulse 84/min., regular, small volume, BP 130/80 mm. Hg, large 'a' wave in JVP. Cardiac impulse in 5th space in left midclavicular line, Right ventricular in type, Left parasternal lift. Ejection click with mid systolic murmur at pulmonary area, fixed splitting of second sound, early diastolic murmur.

Basal crepitations; Liver not palpable.

Investigations—ECG: RAD vertical heart and clockwise rotation, marked right ventricular hypertension. Teleradiogram: (Fig. 1) Enlarged right ventricle. Pulmonary arteries grossly enlarged proximally but poor vasculature peripherally.

Investigations done at Hammersmith Hospital in January/February, 1960 (through the courtesy of Dr. J. F. Goodwin).

Cardiac catheterisation—

<table>
<thead>
<tr>
<th>Site</th>
<th>Saturation</th>
<th>Pressure in mm. Hg.</th>
</tr>
</thead>
<tbody>
<tr>
<td>S.V.C.</td>
<td></td>
<td>54</td>
</tr>
<tr>
<td>R.A.</td>
<td>(high)</td>
<td>a=6</td>
</tr>
<tr>
<td></td>
<td>(mid)</td>
<td>v=6</td>
</tr>
<tr>
<td></td>
<td>(low)</td>
<td>71</td>
</tr>
<tr>
<td>R.V.</td>
<td></td>
<td>72</td>
</tr>
<tr>
<td>P.A.</td>
<td></td>
<td>90/5</td>
</tr>
<tr>
<td>F.A.</td>
<td></td>
<td>65</td>
</tr>
<tr>
<td>L.A.</td>
<td></td>
<td>90/55</td>
</tr>
<tr>
<td></td>
<td></td>
<td>a=6</td>
</tr>
</tbody>
</table>

Systemic flow = 3.1; Pulmonary artery flow = 3.0; Left to right shunt = 0.8; Right to left shunt = 0.9; Pulmonary arteriolar resistance 17 units.

Left atrial angiocardiogram demonstrated free flow from left to right atrium. The RV and PA were outlined immediately after the aorta. Extremely small peripheral lung vessels.

Diagnosis—Atrial septal defect with reversed shunt (Eisenmenger Syndrome).

Progress. This patient deliberately became pregnant and was determined to go through it, against the warning by Hammersmith Hospital. Termination was therefore not considered. She was given digoxin, diuretics and trinitrin. She had earlier during her stay in England, been on anticoagulants (phenindione), but these had been discontinued prior to returning home.

On 16.8.61 after complaining of haemoptysis for two days, she was admitted to hospital, put to bed and the digoxin and diuretic therapy increased. She discharged herself on 18.8.61 as she had no further haemoptysis. At that time she showed no significant change in cardiovascular signs though she seemed slightly breathless at rest.

On 13.9.61 (28 weeks pregnant), no change was noticed. She was allowed to travel 100 miles to join her husband but was advised to reduce exertion and excitement to a minimum, and to continue therapy.
On 11.11.61 she was admitted to the Castle Street Hospital for Women, with labour pains. There did not appear to be any undue cardiac distress but digoxin, 0.5 mg. i.m. was given. She delivered a live normal male infant weighing 5 lbs. 1½ oz. (2.3 kg.) after an episiotomy under local anaesthesia. Labour lasted six and a half hours; there was no cardiac distress or increase in cyanosis at any time. She was got out of bed on the second day and discharged on 11.11.61. She breast-fed the baby for six weeks and was able to look after it with help. She developed angina partially relieved with trinitrin. She was advised to take an oral contraceptive drug and was given "Conovid". She developed mild ankle oedema, which was relieved by taking a diuretic. On 28.11.62, she reported minor haemoptyses in the previous three weeks. She was looked after her baby herself. Last seen on 27.2.63, of all drugs, occasional haemoptyses, but otherwise felt well.

Discussion
A study of the literature revealed a case comparable in some respects to ours. Kirklin, Wedman, Burroughs, Burchell and Wood (1956) in a paper on the correction of ASD, mention one with the following features: ASD, partial anomalous pulmonary venous connection, a bidirectional shunt with severe pulmonary hypertension (PA pressure S/D 86/33 mm. Hg) who was pregnant at the time of the operation. When studied two weeks after the operation she had a residual right-to-left shunt of 20 per cent (preoperatively 25 per cent). This patient was not cyanosed (radial artery oxygen saturation 94 per cent). She subsequently had a normal delivery.

It should be noted that surgery (for closure of defect) was contra-indicated in our patient as she had a pulmonary vascular resistance of 17 units (1360 dynes/sec/cm²), and a pulmonary blood flow which was less than the systemic flow (3.0 and 3.1 L/min. respectively). A pulmonary vascular resistance exceeding 12 units (960 dynes/sec/cm²) and pulmonary blood flow less than 1.75 times systemic flow are contra-indications to closure of these defects (Wood 1958).

The presence of persistent cyanosis has been regarded as a contra-indication to pregnancy (Taussig, 1960). Mendelson and Pardee (1941) believe that cyanosis per se is not a contra-indication.

Burwell and Metcalfe (1958) point out that the reduction of peripheral resistance which occurs in normal patients during pregnancy or the development of peripheral vascular 'shock', would increase the shunt in a patient with an abnormal communication between the heart chambers. Thus patients with a right-to-left shunt and cyanosis are liable to syncope, increased cyanosis and even death during delivery and immediately post partum. They advocate prevention of blood loss. Though our patient gave a history of syncope on exertion during childhood no tendency for this was noticed during pregnancy, at delivery or during the puerperium. Blood loss at delivery was estimated at 4 ozs. (112 ml) before expulsion of the placenta.

The decision to withhold anticoagulants during this patient's pregnancy merits discussion.

Profuse haemoptysis was a cause of death in 29 per cent of 42 fatal cases of the Eisenmenger syndrome (Wood, 1958). Necropsy proved that the cause of the haemoptysis was usually pulmonary infarction from arterial thrombosis—this was not established in every case, however. Edwards (1957) suggested that in this type of case dilation of the small arteries and arterioles reached extreme proportions with aneurysm-like formation. Where these dilated vessels lay near airspaces they might herniate into them and rupture to cause haemoptysis. Our patient's haemoptyses were probably due to rupture of small arterioles. She had several; had each of these been due to pulmonary infarction, there would have been some clinical deterioration—such as increasing cyanosis or dyspnea. This was not seen either during or after a haemoptysis.

It was also thought that anticoagulants were dangerous to the fetus.

This patient was unwilling to be surgically sterilised and she was therefore given Norethynodrel (Conovid). Transient ankle oedema was noticed, responding to an oral diuretic. There was no evidence of cardiac failure and oedema was thought to be due to the salt-retaining effect of the
progestational steroid; it did not recur. Sodium retention may be noted in cardiac patients given these drugs; they do cause this in normal females but only in the first cycles of administration.

Summary
A successful pregnancy in a patient with Eisenmenger's syndrome (ASD with shunt reversal) is described. Certain special problems during the pregnancy have been discussed.

We wish to express our thanks to Prof. J. F. Goodwin, Consultant Physician, Postgraduate Medical School of London, for details of investigations carried out on this patient when she was under his care and for help with the references.

REFERENCES
EDWARDS, J. E. (1957): Functional Pathology of the Pulmonary Vascular Tree in Congenital Cardiac Disease, Circulation, 15, 164.
—- (1958): The Eisenmenger Syndrome, ibid, ii, 701, 755.

SUBDURAL HÆMATOMA
A. DAS GUPTA, M.R.C.P., Ph.D.
Medical Registrar St. Mary's Hospital, Newport, Isle of Wight.

Since the original report on extradural hæmorrhage by Pott (1760) and the review by Jacobson (1885) there have been numerous reports in the literature of this disorder in its various clinical forms (see McKissock, Richardson and Bloom, 1960). The incidence of subdural hæmatoma, following head injury, according to Cecil and Loeb's "Text book of Medicine," is variably estimated at 1 to 10 per cent. It is classified as acute or chronic according to whether the symptoms develop at the time of injury or appear after an interval of weeks or months. In some of the latter cases the previous head injury may have been so slight as to be forgotten. The most delayed case in the literature (to my knowledge) was reported by Grant (1944) who suggested that an anterior cranial fossa hæmatoma originated 6 years prior to surgery.

"The old adage that subdural hæmatoma is never found when it is diagnosed and is always found when it is not diagnosed, should not be true today if a history of head injury is forthcoming" (Northcroft, 1962). But when there is no such history the problem of diagnosis becomes much more difficult. Subdural hæmatoma should always be borne in mind in the differential diagnosis when an intracranial space occupying lesion is suspected.

Case History
A 16-year-old boy was admitted on May 20th, 1962, with a history of persistent frontal headache for three weeks. There was one episode of vomiting. For the past five months prior to admission he had had intermittent blurred vision. No history of any head injury. No previous illness of any significance.

On examination, he was alert and well orientated, there was a mild neck stiffness though Kernig's sign was negative. Bilateral papilledema was found (left more than right). There was bilateral first degree nystagmus and a mild left upper motor neurone facial palsy. In the extremities there was a fine tremor on the left with mild adiadochokinesis. Slightly increased reflexes on the left side. Planters were bilaterally flexor. After much hesitation, lumbar puncture was done which showed increased pressure but otherwise normal CSF. Subtentorial tumour, (possibly mid-line cerebellar), was diagnosed and the boy was transferred to the neurosurgical unit of a London hospital. The boy was taken to the operating theatre on May 25th, 1962, with a view to ventriculography. Biparietal burrholes were placed and much to the surprise of the surgeons, bilateral subdural hygromas were found. Over 100 ml. of fluid were aspirated on the first occasion. Subsequently subdural tapping resulted in markedly decreased amounts of fluid. The patient remained alert although he continued to have mild headaches. The neurological signs remained more or less the same. He was transferred to a convalescent hospital.

After transfer to the latter hospital he started complaining of increasing headache and appeared to be somewhat drowsy. He therefore was re-admitted to the same neurosurgical unit on June 5th, 1962.
Successful Pregnancy in a Patient with Eisenmenger's Syndrome: Atrial Septal Defect with Shunt Reversal
S. H. P. Nanayakkara and E. V. Pieris

Postgrad Med J 1964 40: 670-672
doi: 10.1136/pgmj.40.469.670

Updated information and services can be found at:
http://pmj.bmj.com/content/40/469/670.citation

These include:

Email alerting service
Receive free email alerts when new articles cite this article. Sign up in the box at the top right corner of the online article.

Notes

To request permissions go to:
http://group.bmj.com/group/rights-licensing/permissions

To order reprints go to:
http://journals.bmj.com/cgi/reprintform

To subscribe to BMJ go to:
http://group.bmj.com/subscribe/