Eisenmenger’s syndrome in pregnancy: maternal and fetal mortality in the 1990s

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To determine maternal and fetal mortality associated with Eisenmenger’s syndrome in the UK, a postal questionnaire was sent to 225 NHS obstetric units with neonatal intensive care units, requesting information about maternal and fetal outcome in cases of Eisenmenger’s Syndrome between 1991 and 1995. Fifteen cases were identified. The maternal mortality was 40% and fetal loss 8%. Only 15% of infants were born at term. Maternal mortality associated with Eisenmenger’s syndrome remains as high as it has been for the past 50 years. Pooling of national data on rare medical conditions in pregnancy is required to aid management of individual cases.

Introduction

In each of the last four triennial Confidential Enquiries into Maternal Deaths in the United Kingdom there have been between six and ten maternal deaths associated with congenital heart disease reported, representing between 1.9 and 3.8% of all maternal deaths1. During this period nine of the deaths were associated with Eisenmenger’s syndrome, defined as pulmonary hypertension secondary to an uncorrected left-to-right shunt from a ventricular septal defect, atrial septal defect, or patent ductus arteriosus2. Because of these structural defects, pulmonary flood flow is increased in childhood, leading to fibrosis in the pulmonary vasculature and eventually, the development of pulmonary hypertension. This leads to the shunt reversing and the passage of blood directly from the right side of the heart to the systemic vasculature, which produces increasing cyanosis. The development of pulmonary hypertension in such patients renders the underlying defect inoperable. Heart and lung transplantation is then the only surgical option3.

Case reports and reviews spanning the past 50 years suggest that maternal mortality and perinatal mortality is high in Eisenmenger’s syndrome and is usually quoted at about 50%, although such figures are of uncertain validity. This is because most papers report personal series over many years and give little idea whether new strategies for management, such as the use of selective pulmonary vasodilators, might have affected the outcome. We therefore carried out a national survey to investigate the mortality of Eisenmenger’s syndrome in pregnancy within the past five years, in an attempt to obtain a more up to date picture than is currently available from the literature. We considered that this information would be of use for counselling women with Eisenmenger’s syndrome who are considering pregnancy and for planning their care, should they become pregnant.

Methods

A questionnaire was sent to all NHS obstetric units in the UK identified as having neonatal intensive care units in the Directory of Emergency and Special Care Units 1996 (CMA Medical Data, Cambridge 1996). Information about any pregnant women with Eisenmenger’s syndrome cared for in the unit within the previous five year period (1991–1995) was requested. Only the outcome for mother and baby and the gestational age at delivery were recorded. A further copy was sent if no reply was received within two months. Only those cases in which the pregnancy extended into the second trimester were analysed.

Results

Two hundred and twenty-five questionnaires were sent; overall response rate was 192 replies. Fifteen cases of Eisenmenger’s syndrome were reported in which the pregnancy continued into the second trimester of pregnancy; six women (40%) and one infant (8%) did not survive the pregnancy. The median gestation at birth was 36 weeks (range 20–40 weeks). The median duration of pregnancy of mothers who survived was no different to that of those who died: 36 weeks (range 20–40 weeks) compared with 36 weeks (range 26–36 weeks), respectively (Mann–Whitney rank sum test, \( P = 0.9 \)). Eleven out of the 13 babies for whom information was available were recorded as born before 37 completed weeks of
gestation, an 85% preterm birth rate. The only infant which did not survive was born before 24 weeks. All the infants were grossly normal at discharge, apart from one born at 26 weeks gestation.

Discussion

Women with Eisenmenger's syndrome who become pregnant are rare. The review by Gleicher et al.\(^4\) of the literature between 1948 and 1978 identified 44 published cases and a similar review by Stoddart and O'Sullivan\(^5\) of more recent literature (1979–1991) identified 30 published cases. Alvira et al.\(^6\) prospectively studied 12 women between 1987 and 1993. A 81% response rate to our questionnaire covering a five-year period identified only 15 cases. Although there may have been under-reporting, we feel this is unlikely since cases of Eisenmenger's syndrome tend not to be forgotten, and we deliberately chose a relatively short time period for our survey. For the same reason, the 19% of units which did not respond are less likely to have had cases.

There is agreement in the literature that pregnancy is poorly tolerated in Eisenmenger's syndrome. Furthermore, the successful management of one pregnancy does not imply a similar outcome in subsequent ones, since the death rate for second or third pregnancies is probably no different from that of the first\(^4\), as there may well have been deterioration in the mother's condition in the interim.

Gleicher et al.\(^4\) found that maternal mortality per pregnancy was 30%, but that 52% of women with Eisenmenger's syndrome who had ever been pregnant, died during pregnancy or the puerperium because of the added risk of more than one pregnancy. Likewise Stoddart and O'Sullivan\(^5\) recorded a mortality rate per pregnancy of 26% but 39% of women died. Morgan Jones and Howitt\(^6\) reported rates of 27% and 31% and Alvira et al.\(^6\) reported rates of 30 and 33%, respectively. We did not request information about parity, but the 40% maternal mortality rate in our study accords with these figures for overall mortality. The perinatal mortality rate was in contrast surprisingly low, presumably because of advances in neonatal care in recent years. It is of particular concern that five babies survived but were left motherless.

We restricted the information requested in the questionnaire deliberately. Although details of mode of delivery or the underlying cause of Eisenmenger's syndrome, for example, would be of interest, we felt a request for a greater amount of information would have reduced the response rate and our main concern was the mortality rate.

It appears that the prognosis for the woman with Eisenmenger's syndrome who becomes pregnant has not improved over the past 50 years. This may be because more (and therefore proportionately sicker) women are surviving; there is evidence to support this view from recent surveys\(^8\). The incidence of Eisenmenger's syndrome amongst women of reproductive age and the pregnancy rate amongst this group remains to be determined. The small numbers of our survey do not allow detailed analysis or extrapolation, but it represents the first attempt to gain national data about a rare and serious cardiac condition. From an anaesthetic viewpoint particularly, pooling of national data on cardiac disease in pregnancy has considerable potential for aiding management of individual cases. The initiative has been taken to create a national high risk registry of obstetric anaesthesia relying on voluntary reporting of cardiorespiratory cases\(^9\). It is planned to extend this to include other rare conditions in pregnancy.

References


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