CARDIOMYOPATHY OF PREGNANCY AND PUERPERIUM

(A report of Two cases with Review of Literature)

by

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The diagnosis of cardiomyopathy is considered for those cases of heart failure which come for the first time during the last trimester of pregnancy or during the puerperium and where there is no past history, symptoms or physical findings of a similar episode and when the known causes of heart failure such as toxaemia, hypertension, valvular disease of the heart, coronary artery disease, anaemia, etc. can be reasonably excluded (Walsh *et al* 1965 Middleton 1964). Such cases have been known in the literature for over fifty years, but there has been a better recognition of this condition in recent years and now this condition is considered as a specific clinical entity in any standard classification of cardiomyopathies. It is not possible to give the exact incidence of this condition, as many mild cases go unrecognised and unreported and of the serious cases which seek admission to the hospitals all are not documented. However, some authors (Pierce *et al* 1968; Meadows 1957) have given its incidence as one in 1300 deliveries or one in 4000 hospital admissions which from our own experience seems to be pretty high.

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Below are presented two cases which came under our observation in this institution.

Case 1.

Mrs. S. K., 25, primipara was admitted on 5-8-1970 for palpitation, breathlessness on exertion (grade II), pain in the chest and pain in the right side of the abdomen since her last illness about 1½ years back. An enquiry into the past illness revealed that she had developed breathlessness on exertion and later generalised oedema about 1½ years back when she was eight months pregnant. She was diagnosed and treated as a case of congestive heart failure at that time by a private practitioner. X-rays taken at that time showed the heart to be enlarged and there was congestion in the lungs. There was no history of breathlessness, rheumatic arthritis or oedema prior to her last conception.

On examination, the patient was well nourished and fairly built without any cyanosis, oedema or anaemia. Pulse rate was 100 per minute regular and of fair volume and was of pulsus alterans type. Blood pressure was 110/70. Apx beat was diffusely located in the 5th intercostal space outside the midclavicular line. Both the heart sounds were faint and distant. No murmur or rub could be heard over the precordium. Liver was not plapable but there was diffuse tenderness in the right hypochondrium. Jugular venous pulsations were normal. Lungs were clinically normal.

E.C.G. showed flattening and inversion of T-waves in the chest and precordial leads. X-rays of the chest showed the heart to be

enlarged. Other investigations were, Hb 11 gm per 100 cc of blood. E.S.R. was 70 mm in first hour. Urine was normal.

Case 2.

Mrs. N., 33, 5th para, developd cough and breathlessness on exertion when she was 8½ months pregnant. She delivered at home, full-term, 12 days prior to the admission. She complained of breathlessness (grade III) and cough with expectoration and slight oedema over the feet. There was no history of similar episodes or rheumatic arthritis in the past.

On examination, she was a moderatly built woman without any obvious anaemia, cyanosis or oedema. Pulse rate was 120 per minute and regular. Blood pressure was 100/70 mm of Hg. Heart was enlarged clinically. Both the heart sounds were clear. No murmur or a rub ciuld be heard over the precordium. There was gallop rhythm over the precordium. Lungs showed bilateral rhonchi and occasional crepitations. Liver was enlarged 3 fingers below the costal margin and acutely tender. X-rays confirmed enlargement of the heart and E.C.G. showed flattening of the T-waves and depression of ST segment in L1, L2, AVF and V5 and V6 leads. Other investigations were within normal limits. She was diagnosed as a case of congestive heart failure and put on the usual treatment of back-rest, digoxin, diuretics and antibiotics. There was some improvement in breathlessness and cough, but the pulse rate remained persistently high. E.C.G. examination, 7 months later, did not show any appreciable improvement in the findings, though she had clinically improved on the maintenance therapy of digoxin and diuretics during this period.

Comments and discussion

Predisposing Factors: (1) Race: Almost all the earlier reports have described this condition among the negro women (Walsh *et al,* 1965; Meadows, 1957; Gilchrist, 1967; Stuart, 1968) giving a bias that probably this condition is peculiar to negro women and that race has something to do for its causation. But, in recent years the condition has been commented upon from all parts of the world—U.K. (Bridgen, 1957; Gilchrist, 1963) Saudi Arabia (Perrine, 1967), South Africa (Seftel and Sussex, 1961), Japan (Sakakibara *et al,* 1970) and China as quoted by Perrine. It appears from this that the disease is not peculiar to any particular race.

(2) *Age, Parity, and Lactation:* Most of the authors agree that this condition is more common in the higher age group. Dominant age in one series was 25-35 years (Meadows), though Johnson *et al* (1966) have described a case in a 14-year old negro woman. Both of our patients were in the age group of 25-30 years.

This condition is said to be more common after multiple pregnancies, pregnancies in rapid succession and after a prolonged period of lactation (Stuart, 1968, Seftel and Sussex, 1961), though cases have been described in primiparae (Johnson *et al* 1966). Case 1 of ours was also a primipara.

(3) *Nutritional and socio-economic status:* Most of the patients mentioned in the literature are said to have come from low socio-economic strata of society (Walsh, Meadows, Perrine, Seftel and Sussex). Nutritional status of these patients have been commented upon by some of these authors very carefully. None of these authors has mentioned any clinical evidence of nutritional deficiency in these patients. Both of our patients were fairly nourished and were without any clinical evidence of nutritional deficiency.

The views as regards the exact nature of the disease are sharply divided. Some authors believe that the cardiomegaly of unknown origin or of familial origin (Brown *et al* 1967) is already existing in such patients and that pregnancy super-

imposing merely acts as a trigger and unmasks it and the heart fails due to normal circulatory haemodynamic changes of pregnancy. On this hypothesis one should expect majority of cases occurring immediately after the delivery when the circulatory load is maximum and not weeks and months afterwards when the haemodynamic changes have settled down to normal. Moreover, familial cases of cardiomyopathy rarely. if ever, fail during pregnancy or puerperium (Brown *et al*).

Others believe that there is no disease entity as such of postpartum heart disease and that it is merely a complex of symptoms due to a variety of causes which might have damaged the heart and caused heart failure (Benchimol *et al*, 1959; Bashour and Winchell, 1954). These authors emphasise that careful analysis of history, findings and investigations of such cases to find a cause should be made. Benchimol *et al* could find one or the other cause in 17 out of 18 patients with postpartum heart failure and in only one case the diagnosis was suggestive of cardiomyopathy of pregnancy and puerperium. Among the causes listed were toxaemias (11 cases), pre-existent hypertension (3 cases), Chaga's disease 1 case), miliary tuberculosis (1 case) and nutritional cause (1 case). Similarly, one of the two cases described by Bashur and Winchell was believed to be that of influenzal myocarditis and in the other case the heart was believed to have been damaged by hypersensitivity reaction to sulphonamide. As a matter of fact, hypersensitivity reactions to a wide variety of drugs and even to the products of conception (Douglas *et al*, 1959) which have been found circulating in the blood stream have been invoked to explain the damage to the heart in such cases.

The majority, however, are of the opinion that this is a specific condition and that pregnancy and puerperium are its immediate causes. How exactly this condition is caused is not known. It is probable that multiple pregnancies, frequent pregnancies and prolonged period of lactations adversely affect the myocardium by altering its metabolism or these very conditions lower the nutritional status of the pregnant women who mostly belong to the low socio-economic strata of society and these factors acting collectively aggravate the subclinical nutritional deficiency which might be already existing in them.

Clinical features: Barring its association with gestation, the clinical features, E.C.G. findings and radiological features of this condition are not specific and are the same in congestive heart failure due to cardiomyopathy of any origin.

Meadows (1957) and many other authors have associated this condition predominantly to puerperium though this is also known to occur during the third trimester of pregnancy (Case 1 and Case 2). The onset may occur at any time in the puerperium from 1st day to 50th week afterwards (Perrine), though majority of cases occur around 20th week. Onset in most cases is insiduous and mild cases may not be diagnosed. A case of fulminating myocarditis with an acute onset has also been described by Faraque, (1965). The patient commonly presents as any other case of congestive heart failure with breathlessness on exertion, orthopnoea, nocturnal dyspnoea, cough and fatigue and on examination enlarged tender liver, engorged jugulars and dependent or generalised oedema. There is persistent tachycardia which may be regular or irregular. Case 1 had pulsus alternans. Heart may or may not be enlarged clinically. Heart sounds may be

normal or be muffled and distant as in Case 1. There may be insignificant grade I or grade II systolic murmur over the praecordium. There may be gallop rhythm as in Case 2. There is commonly no evidence of any valvular lesion, anaemia, hypertension, toxaemia or any other disease to account for the heart failure.

E.C.G. findings, as has been pointed out, are not specific and various types of conduction defects and arrythmias and various combinations of ST segment depression and flattening and inversion of the T waves have been described. Both of our cases showed flat or inverted T-waves in the left sided precordial leads. Radiological findings are also non-specific and consist of enlargement of heart and congestion in the lungs. The course of the disease is protracted. The case may recover completely or may recur in subsequent pregnancies. Mortality rate is said to be between 20% and 60% (Walsh *et al*, 1965).

Pathological features: Johnson *et al* (1966) have reviewed postmortem findings in 26 fatal cases reported in the literature and added one of their own. As with clinical features, naked eye and microscopic findings are not specific The heart is enlarged and its chambers dilated with many mural thrombi over the endocardium. Microscopic picture shows the degeneration of the muscle fibres consisting of cloudy swelling, fragmentation of fibres and loss of striations and pyknosis of the nucleii. There is cellular infiltration mostly consisting of lymphocytes, mononuclear cells and occasionally macrophages and an attempt at reparative process as evidenced by endocardial thickening and fibrosis.

Endomyocardial biopsy techniques are being applied to elucidate the nature of the disease. Sakakibara *et al*, (1970) have studied the ultrastructure in such biopsy specimens by histochemical methods. They found a specific peculiar proteineceous substance that stained negatively for mucopolysachharide. The importance of this finding remains to be elaborated.

Management: Management of these cases is on conservative lines as in, congestive heart failure due to any cause and consists of rest, digoxin, diuretics, sedatives and salt restricted diet. The results are not very encouraging and the improvement is slow (Case 2). Heart failure may recur in subsequent pregnancies and it may be advisable to advise against future pregnancies, though the consensus of the opinion is not clear on this (Stuart, 1968).

Summary

Two cases of cardiomyopathy of pregnancy and puerperium have been presented and the literature on the subject is reviewed. The condition is rare and has been reported from all the regions of the world. This condition is now regarded as a specific clinical entity precipitated by pregnancy and puerperium though exact factors responsible for this remain unknown. Management of such cases is on conservative lines; though improvement on treatment is slow, the course is protracted and there is tendency to recur in subsequent pregnancies.

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