DOI: 10.7860/JCDR/2014/9924.4803

Original Article

Obstetrics and Gynaecology Section

Are We Missing Hypertrophic Cardiomyopathy in Pregnancy? Experience of a Tertiary Care Hospital

POOJA SIKKA[1], VANITA SURI[2], NEELAM AGGARWAL[3], SEEMA CHOPRA[4], AJAY BAHL[5], RAJESH VIJAYVERGHIA[6]

ABSTRACT

Background: Controversies persist regarding risks associated with pregnancy and delivery in women with hypertrophic cardiomyopathy (HCM). To date, pregnancy outcome data for these patients is scarce. We report the experience of pregnancies with HCM in a tertiary care hospital.

Materials and Methods: Data regarding cardiac illness and obstetric profile of all women attending the cardio-obstetrics clinic from January 1990 to December 2012 were studied. The records of cardiac illness of all women were checked and all patients with HCM were included in the study.

Results: Out of total 2016 patients booked in the cardio-obstetrics clinic between 1990 and 2012, only 4 women were found to have a diagnosis of HCM (0.2%). Of these, 2 women with left ventricular outflow tract obstruction and one with non-obstructive HCM had only mild symptoms and tolerated pregnancy and labour well. One patient had HCM with restrictive physiology developed heart failure and intra-uterine fetal death.

Conclusion: HCM is underdiagnosed and rarely identified in pregnancy. Most patients with HCM tolerated pregnancy well, howeverone patient with restrictive physiology developed heart failure during her first pregnancy.

Keywords: Hypertrophic cardiomyopathy, Pregnancy

INTRODUCTION

Hypertrophic cardiomyopathy (HCM) is a complex, genetic cardiac disorder, the exact incidence of which in pregnancies is unknown. Both obstetricians and cardiologists face the difficult issue of managing HCM in pregnancy as controversies still persist regarding risks associated with pregnancy and delivery. HCM is considered a WHO class 2 and 3 risk, implying thereby that there is a moderate risk of morbidity in most women and a significant risk for few [1]. To date, pregnancy outcome data for these patients is scarce [2]. Though small studies have reported no deaths associated with pregnancy in HCM, some have highlighted several maternal complications [3]. Widespread use of echocardiography and both genetic and clinical screening of families with HCM has led to a higher number of women being diagnosed as HCM nowadays.

We report the experience of pregnancies with HCM in our hospital which is a tertiary care hospital in Northern India.

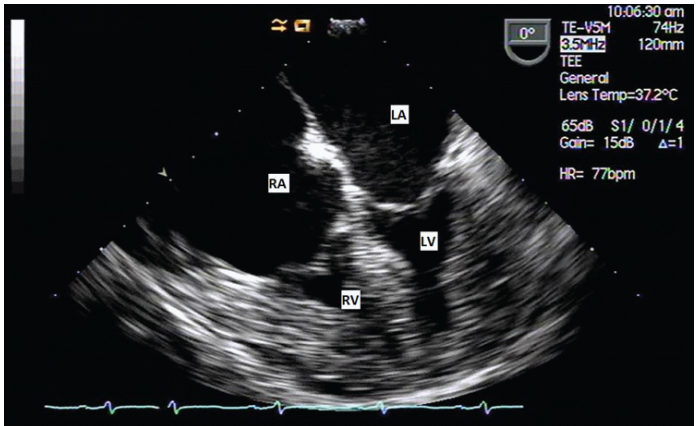
MATERIALS AND METHODS

Data regarding cardiac illness and obstetric profile of all women attending the cardio-obstetrics clinic at the Post-graduate Institute of Medical Education and Research, Chandigarh, India from January 1990 to December 2012 were studied. The records of cardiac illness of all women were checked and all patients with HCM were included in the study. The criteria for diagnosis of HCM was unexplained left ventricle hypertrophy greater than or equal to 13 mm or > 2 standard deviations corrected for body surface area by echocardiography [4]. Patients with systemic hypertension, aortic stenosis or any other systemic or cardiac disease capable of producing the magnitude of wall thickening were excluded.

RESULTS

Out of total 2016 patients booked in the cardio-obstetrics clinic at our centre between 1990 and 2012, four women were found to have a diagnosis of HCM. Total number of pregnancies in these four women was eight. Four deliveries had occurred in our institute and other four were previous outside deliveries. The case summaries of the four women are described below.

Case 1: This 32-year-old lady, a known case of HCM was referred at 28 wk of gestation with history of palpitations and breathlessness on exertion (NYHA class II). Echocardiography revealed HCM with left ventricular outflow tract obstruction (LVOTO). Septal thickness was 20mm and she had a severe LVOTO with a peak systolic gradient of 76 mmHg. She was started on metoprolol and there was no further worsening of symptoms. She went into spontaneous labour at 37 wk and delivered a healthy baby. Her previous pregnancy was five years back. She had complained of chest pain during her first pregnancy and on evaluation was diagnosed to have HCM. She had delivered vaginally, uneventfully in another hospital. She remained asymptomatic after her second delivery for four years. She then



[Table/Fig-1]: Transoesophageal echocardiography of patient with hypertrophic cardiomyopathy with restrictive physiology (case 4) showing biventricular hypertrophy with biatrial enlargement. Biatrial enlargement is suggestive of restrictive physiology

developed dyspnoea and angina on exertion that worsened to NYHA class III. Alcohol septal ablation was carried out in our institute six years after her second delivery with partial relief of symptoms. An implantable cardioverter-defibrillator was also implanted due to high risk of sudden death. A second alcohol ablation was carried out three years later for residual gradient. She has completed 10 y follow up after her last delivery and has only mild breathlessness on heavy exertion. She does not have any left ventricular outflow tract gradient.

Case 2: This 35-year-old lady, not a known case of HCM was referred in her third pregnancy at 32 wk of gestation with history of palpitations and dyspnoea (NYHA class II). Cardiovascular system examination was normal and there was no murmur. Echocardiography was suggestive of non-obstructive type HCM, septal thickness of 18mm with mild mitral regurgitation. She was started on metoprolol with significant improvement in symptoms. Her pregnancy continued uneventfully till 39 wk of gestation. She went into spontaneous labour and delivered a healthy baby vaginally. She was asymptomatic in previous two pregnancies time of previous pregnancies which were uncomplicated and she had delivered vaginally in another hospital.

Case 3: This 24-year-old lady was referred at 24 wk of gestation with history of dyspnoea and chest pain (NYHA class II). Clinical examination did not reveal any abnormality. Echocardiography showed HCM with maximal septal thickness of 21 mm with LVOTO. There was gradual improvement in symptoms on bed rest and metoprolol. She went into spontaneous labour at 36 wk and delivered a healthy baby. In her previous pregnancy one year back she was under follow-up at another hospital, had dyspnoea at 26-28 wk, went into preterm labour and delivered a premature baby of 800 g who died on day one of life. Following delivery there was improvement in dyspnoea so she did not undergo any cardiac evaluation.

Case 4: This 28-year-old primigravida was referred to our institute with symptoms of breathlessness at rest, orthopnoea and palpitations. She was previously unaware of having any heart disease. She became symptomatic as early as 24 wk of gestation. Her symptoms rapidly progressed and by 26 wk she went into heart failure with pulmonary oedema and pericardial effusion (NYHA class IV). She was found to have HCM with restrictive physiology [Table/Fig-1]. Maximal left ventricular wall thickness was 19 mm. She had an intrauterine fetal death at this time. She was managed with beta blockers and diuretics. Induction of labour was done with pitocin for pregnancy termination at 28 wk. She delivered uneventfully and there was a gradual improvement in her symptoms after delivery. She has a persistently raised jugular venous pressure and pericardial effusion at 6 y follow up. She is able to carry out usual household activities.

DISCUSSION

Our retrospective investigation of 22 y in pregnant women with underlying cardiac disease revealed only four women having HCM. The incidence of cardiac disorders in pregnancy in our institute is about 3%. The prevalence of HCM in the general population is around 1 in 500 and the same prevalence is expected to be seen in pregnant patients [4]. However, in our review the incidence was 0.2% among cardio-obstetric patients. Such low incidence can only be explained by the supposition that the disease does not become troublesome during pregnancy and thus goes undiagnosed. However, as routine echo in asymptomatic patients is not done in our institute and the facility for genetic screening also does not exist we might be missing the diagnosis of HCM in quite a few asymptomatic women. Other studies have also found a low prevalence of HCM among pregnant women. It was found to be 0.015% in one of the cohort studies on pregnancy with cardiac disease [5].

In our study, 3 out of 4 women had only mild symptoms in the form of palpitations and dyspnoea that improved with beta-blockers. Two of these had LVOTO and one had non-obstructive HCM .Only one pregnancy ended preterm but it is likely that it was not HCM related since she had only mild symptoms. Few data suggest that one of the risk factors for clinical deterioration during pregnancy is presence of LVOTO [6]. However, theoretically, any increase in preload which occurs in conditions like pregnancy will reduce the obstruction and there should be no worsening of symptoms [7]. In Avila's study there was a small but significant increasein LVOTO gradient in pregnancy compared with pre-pregnancy, but those with outflow obstruction did not have more complications [7]. One retrospective study also found that LVOTO obstruction was not a predictor of complications. Though the number of patients in our review was small, LVOTO was well tolerated and did not indicate a risk for worsening during pregnancy.

Only one patient did not tolerate pregnancy and deteriorated rapidly in middle of pregnancy. She had HCM with restrictive physiology. Many studies have shown restrictive physiology to be bad prognostic indicator for various complications like heart failure, arrhythmias and sudden death even in absence of pregnancy [8]. Pregnancy in restrictive type of HCM is unreported. Though, in our study there was only one patient with restrictive physiology,her poor pregnancy outcome suggests that maybe pregnancy is contraindicated in restrictive type of HCM.

All patients in our study received metoprolol, since, there is considerable experience of its use in pregnancy [9]. Two pregnancies ended preterm, the preterm delivery rate being 25% in pregnancy with HCM. None of the babies born at term had intrauterine growth retardation. Elkayam in a review on 82 pregnancies with HCM showed a 20% spontaneous abortion rate and 10% low birth babies [10]. None of the patients was delivered by caesarean. Literature also favours vaginal mode of delivery in patients with HCM, caesarean reserved only for obstetric indications [1]. None of the patients were given epidural analgesia, although not contraindicated, epidural and spinal anaesthesia must be given in these patients with caution due to risks associated with consequent vasodilation [11].

Ideally every woman with HCM should have a pre-pregnancy genetic counselling regarding maternal risks associated with pregnancy and associated risk to the baby as HCM is inherited as autosomal dominant disorder [12]. In our study, all women were referred during pregnancy so none had come for a preconceptional counselling. The patient with restrictive physiology (case-4) has been advised against future pregnancy.

CONCLUSION

HCM is underdiagnosed and rarely identified in pregnancy. Most patients with HCM tolerated pregnancy well, however one patient with restrictive physiology developed heart failure during her first pregnancy.

REFERENCES

[1] Regitz-Zagrosek V, BlomstromLundqvist C, Borghi C, Cifkova R, Ferreira R, Foidart JM, et al. ESC Guidelines on the management of cardiovascular diseases during pregnancy: the Task Force on the Management of Cardiovascular Diseases during Pregnancy of the European Society of Cardiology (ESC).*Eur Heart J.* 2011;32:3147-97.

[2] Krul SP, van der Smagt JJ, van den Berg MP, Sollie KM, Pieper PG, van Spaendonck-Zwarts KY. Systematic review of pregnancy in women with inherited cardiomyopathies. *Eur J Heart Fail.* 2011;13:584-94.

[3] Autore C, Conte MR, Piccininno M, Bernabò P, Bonfiglio G, Bruzzi P, et al. Risk associated withpregnancy in hypertrophic cardiomyopathy. *J Am Coll Cardiol.* 2002;40:1864-69.

[4] Maron BJ, Gardin JM, Gflack JM, Gidding SS, Kurosaki TT, Bild DE. Prevalence of hypertrophic cardiomyopathy in the general population of young adults. Echocardiographic analysis of 4111 subjects in the CARDIA Study. Coronay Artery Risk Development in (Young) Adults. *Circulation*. 1995;92:785-89.

[5] Avila WS, Rossi EG, Ramires JA, Grinberg M, Bortolotto MR, Zugaib M, et al. Pregnancy in patients with heart disease: experience with 1,000 cases. *Clin Cardiol.* 2003;26:135-42.

[6] Thaman R, Varnava A, Hamid MS, Firoozi S, Sachdev B, Condon M, et al. Pregnancy related complicationsin women with hypertrophic cardiomyopathy. *Heart.* 2003;89:752-56.
[7] Avila WS, Amaral FMC, Ramires JA, Rossi EG, Grinberg M, Bortolotto MR, et al. Influence of pregnancyon clinical course and fetal outcome of women with hypertrophic cardio myopathy. *Arq Bras Cardiol.* 2007;88:423-28.
[8] Kubo T, Gimeno JR, Bahl A, Steffensen U, Steffensen M, Osman E, et al. Prevalence, clinical significance, and genetic basis of hypertrophic cardiomyopathy with restrictive phenotype. *J Am Coll Cardiol.* 2007;49:2419-26.
[9] Rubin PC. Beta blockers in pregnancy. *N Engl J Med*.1981;305:1323-36.
[10] Elkayam U, Dave R (1998). Hypertrophic cardiomyopathy and pregnancy. In: Elkayam U, Gleicher N (ed) Cardiac Problems in Pregnancy, *3rd Edn. Wiley-Liss, Inc., New York, pp* 101-109.
[11] Walker D, Kaur N, Bell R, Walker F. Hypertrophic obstructive cardiomyopathyand pregnancy: University College London Hospitalexperience. *Minerva Anestesiol.* 2007;73:485-86.
[12] ICIN working group on Hereditary Heart Diseases. Genetic diagnostics and genetic counselling in Hypertrophic Cardiomyopathy (HCM). *Neth Heart J.* 2010;18:144-59.

PARTICULARS OF CONTRIBUTORS:
1. Assistant Professor, Department of Obstetrics and Gynaecology, Postgraduate Institute of Medical Education and Research, Chandigarh, India.
2. Professor, Department of Obstetrics and Gynaecology,, Postgraduate Institute of Medical Education and Research, Chandigarh, India.
3. Assistant Professor, Department of Obstetrics and Gynaecology, Postgraduate Institute of Medical Education and Research, Chandigarh, India.
4. Assistant Professor, Department of Obstetrics and Gynaecology, Postgraduate Institute of Medical Education and Research, Chandigarh, India.
5. Associate Professor, Department of Cardiology, Postgraduate Institute of Medical Education and Research, Chandigarh, India.
6. Associate Professor, Department of Cardiology, Postgraduate Institute of Medical Education and Research, Chandigarh, India.

NAME, ADDRESS, E-MAIL ID OF THE CORRESPONDING AUTHOR:
Dr. Pooja Sikka,
Assistant Professor, Department of Obstetrics and Gynaecology, Postgraduate Institute of Medical Education and Research, Chandigarh-160012, India.
Phone : 8146553265, E-mail : drpoojasikka@yahoo.com

FINANCIAL OR OTHER COMPETING INTERESTS: None.

Date of Submission: **Jun 16, 2014**
Date of Peer Review: **Jul 31, 2014**
Date of Acceptance: **Aug 02, 2014**
Date of Publishing: **Sep 20, 2014**