BSH Heart Failure Day for Revalidation and Training 2017

Presentation title: Case Presentation - obstetrics

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Conflicts of interest: None
Heart Failure & Pregnancy

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Nil conflict of interest
Heart Disease & Pregnancy

On the increase in the western world due to a number of factors:
  • Increasing age at first pregnancy
  • Increasing prevalence of CV Risk factors – HTN/Obesity/Diabetes
  • Better survival for patients with Congenital Heart Disease

Cardiac disease remains the major cause of maternal death in pregnancy and requires a multidisciplinary approach

Cardiomyopathy – leading cause of these deaths (25%)

Physiological Changes during pregnancy

  30-50% increase in Cardiac Output
  HR increase from 20 weeks onwards
  Heart can dilate
  Hypercoagulability
  Increase in blood volume
  Fall in SVR
Case 1 – Victoria

- First highlighted to local cardiology services due to FHx
- Father was diagnosed with DCM age 45
- Grandmother HF first diagnosed in her 40’s

- Referred to HF Cardiologist at tertiary centre

- Echo: Mild LVSD with Global hypokinesia Low – Normal EF
- MRI Heart: Mildly impaired LV (49%) Mildly impaired RV (47%)

- NYHA Class I
Pregnancy – Age 24yrs

Referred to Cardiologist with expertise in Pregnancy and Heart Disease

1\textsuperscript{st} review at 16 weeks - Asymptomatic, Clinically euvolemic
• Victoria had plans to travel to Lebanon (prearranged)
• Discussed risks and plans for normal delivery – advised against travel

2\textsuperscript{nd} review at 22 weeks - Clinically euvolemic
• Echo Slight deterioration - Moderately Impaired LV EF 40-45%
• Bisoprolol 2.5mg, Referral to HF Nurses

3\textsuperscript{rd} Review 28 weeks – unchanged clinically
• Echo EF 49%
Delivery

Saw anaesthetics at 33 weeks and detailed plans made for delivery

38+3 weeks – induced labour
• Healthy baby girl

Subsequently GP noted beta blocker stopped after delivery and wrote to Cardiologist for advice

• Contraception and future pregnancy plans discussed
• Started back on ACEi (chose not to breast feed)
• Referred back to DGH for follow up (patient choice)
• Genetic screening for family arranged
Points:

• Well managed and co-ordinated care between Cardiology services Locally and at Tertiary centre as well as HF Specialist nurse support

• Integrated care with other teams – Obstetrics and Midwifery as well as Anaesthetics

• Regular reviews and scan (x16 letters from Cardiology over 6 months)

• Proactive approach to management with attempts to mitigate as much risk as possible
Case 2 Sophie

Note: Neither myself nor Dr Barker were involved in this patients care, but the case has been discussed and reviewed within our department.

We have talked to the Consultant involved in her care who is happy for us to present the case.

24 year old female
First presented to DGH with syncope (1st and only episode)

PMH:
Mild learning difficulties
Previous x1 OD attempt – poor coping strategies
Investigations:

• ECG: AF + Pre-excitation - rates 130bpm
• Echo: Mildly impaired LV function ?Thickened posterior wall (LVPWd 14mm)

• MRI: Poor Quality study – Mildly impaired LV, No significant LVH. Study cut short due to metallic artefact in abdomen (dental brace)

• EP Study – Likely Nodo-Ventricular pathway (no ablation)
• Successful TOE Guided Cardioversion
• Bisoprolol and Flecainide stopped
ECG
Echo
MRI

- Mildly dilated LV
- Globally mildly impaired function
- No significant hypertrophy
- Thickened posterior wall on echo is likely a prominent inferolateral papillary muscle
- No Late gadolinium images
6 months later

1st Review at 15 weeks pregnant – (unsure if this was a planned pregnancy)

• X1 light headed episode, otherwise no symptoms
• ECG confirmed AF with rates of 100bpm
• Planned for O/P 24 hour tape, Echo

2nd Review at 22 weeks

• Clinically unchanged
• 24 Hour Tape: AF with rates from 70 – 200bpm, Mean 100bpm
• Echo: Not performed

Management Plan
Management

- Given CHADS2Vasc = 0 → Not for Anticoagulation
- Start Bisoprolol 2.5mg OD for Fast Ventricular Rates
- Review in clinic 6/52

- Letter to obstetricians to co-ordinate care

- No need for urgent cardioversion – planned for DCCV post delivery
5 weeks later – Presents to A&E

• 4/52 hx of cough, SOB, nausea, vomiting, new rash
• Had been treated with Abx from GP and reviewed on 3 occasions (Amoxicillin)

Pulse 200bpm AF, Temp 37.5, BP 100/47, RR 30 Sats 97% RA
Admitting nurse noted she looked “yellow”

Hb 101, WCC 12, CRP 66, Normal U&E, no LFTs, no ABG

On review in A&E:
  • Pale, dry mucous membranes, dehydrated appearance
  • Widespread maculopapular rash over body
  • Chest clear, CVS examination normal

• Treated for ?Gastroenteritis + Drug reaction – Given IV fluids, Cyclizine, Piriton
• Transfer to Maternity Unit
ECG
Further Mx

Following morning → BP 70/40, P 236, Temp 36.7, RR 15, Sats 98%. Given further IV fluids. BP improved to 110 systolic and she reported feeling better.

Afternoon – increasingly SOB, decreased saturations, Pulse 195 → Seen by SHO who felt there were left sided creps

Discussed with a Cardiology SpR who advised to give Bisoprolol

Persistently hypotensive, tachypnoeic, tachycardic. Sats 85%  
Anaesthetics called to ward to review and transferred to High Dependency Unit

Hb 94, WCC 11, U&E normal, ALT 302, Bili 44, Alb 19
pH 7.48, P02 35, PCO2 2.55, BE -8.8, Lactate 7.4

Given further IV fluids and Betablocker. Cardiology contacted again Bedside echo performed.
Bedside Echo
Transferred to Critical Care

Worsening Acidosis (Lactate 12) – Features of Low Output Failure
Decision made to Intubate and Ventilate
CVP inserted – Pressure >20mmHg

• PEA arrest – Adrenaline + DCCV and ROSC

• Emergency Peri-arrest Caesarean Section – Stillborn male child

• Post Delivery – given inotrope but remained cyanosed and hypotensive
  • ABG: pH6.9, PO2 14, pCO2 6, BE -22, Lactate 14

• Decision made to withdraw treatment. Sadly passed away that night.
Referred for Post Mortem

• Congestive Cardiac Failure
• Mild Left Ventricular Hypertrophy
• Mild myocardial fibrosis
• Moderate atheroma in abdominal aorta
• Doliocephaly and High arch palate with crowded teeth,

Sent for molecular autopsy

?Suggestions as to the underlying diagnosis
Danon Disease

- X-Linked genetic disorder with Mutation in LAMP2 gene
- Cardiomyopathy, Skeletal Myopathy, Learning Difficulties
- Classically develops earlier in males with predominantly HCM type cardiomyopathy (90%)
- In females approx. 50% with develop DCM instead.
- Later presentation with less pronounced learning difficulties
- Arrhythmias including pre-excitation (69% males 29% females)
- Possible relation to psychiatric problems with increased prevalence of depression, suicidal ideation, ADHD
Points

• This lady did not receive the care she should have.

• There were multiple failings from other teams in her acute presentation but there are things we could have done?

• Pregnancy is a time when physiology rapidly changes and we need to pre-empt problems and act quickly

• In hindsight perhaps it was not appreciated that the AF and nodo-ventricular pathway were symptoms of an underlying cardiomyopathy rather than the diagnosis itself

Whilst it is important to learn from good practice ➔ it is equally as important to learn from mistakes
What could have been done?

• Pre-pregnancy – no firm diagnosis so missed out on pre-pregnancy counselling.

• During pregnancy - The missed echo was an administrative error - would this have alerted us to a problem sooner?

• Pre-admission – she was left in the community for 4 weeks deteriorating - without being notified to cardiology

• After admission – cardiology were not asked for advice for 24 hours. The severe nature of her condition was not realised till in extremis.
To Think about

Would you have considered this lady as having a cardiomyopathy? Was there enough of a diagnosis to start her on ACEi and discuss pregnancy planning?

Did her learning difficulties mean she was a vulnerable adult and therefore someone we should actively look to monitor for problems?

Do we have a system to ensure administrative errors in higher risk patients (e.g. missed echo) are picked up quickly and acted on?

How can we ensure we are alerted to deterioration in symptoms in the community early?
  • e.g. letter for patient and GP with red flag symptoms
  • Involve cardiac nurses as someone who can keep in touch by phone and ask about symptoms

What can we do to educate colleagues in other departments?
Women with a known history of cardiac disease must be referred for care in a maternity unit where there is a joint obstetric/cardiology clinic or a cardiologist with expertise in the care of women with heart disease in pregnancy.

Women with pre-existing cardiomyopathy of whatever origin must, like all women with heart disease, should have expert involvement and informed pre-pregnancy counselling.

Missing pulmonary oedema and missing acidosis are common mistakes in maternal cardiac deaths.
Personal Experience of (Wife’s!) Pregnancy

Epilepsy is another condition that requires pre-pregnancy counselling – there is a much larger cohort of young women.

On every visit pregnancy and contraception are discussed → My wife has had a clear plan for pregnancy from the age of 18

Hospital appointments:

• 4 obstetricians in 3 hospitals
• 2 epilepsy specialist nurses in 2 separate hospitals
• 2 neurologists in 2 other hospitals
• 1 haematology review
• Multiple Midwife appointments in 2 hospitals + GP surgery
• GP reviews
• X7 USS scans
• Home reviews, antenatal appointments

• On top of all the usual preparations when there are medical issues it can be easy to miss things
• Anything we can do to watch out and ease the burden will help ensure safe pregnancy’s