DMD Care UK Q+A from 10th Jan 2023: Clinician focus

Cardiac care – Dr John Bourke

Question 1: Is it important to always be on the doses of cardiac drugs stated in the guidelines?

Usually yes. However, if the resting heart rate is already well controlled (ie: 75-85 beats/min) at a lower dose, there is usually no need to increases to the 'maximum tolerated dose'. The optimum dose can be reviewed and individualised at subsequent at later follow-ups. Some boys may remain permanently on a lower-than-average dose.

If a patient is on an ACE inhibitor and not drinking sufficient fluids or is hypotensive for other reasons, kidney function may become compromised. In these situations, the dose of ACEi may need to be reduced temporarily but needs to be increased again after recovery.

Question 2: Becker patients and female carriers are recommended to have 3-year cardiac reviews. This can be difficult to arrange because of current NHS booking policies. Often, carriers do not have a specialist clinician to arrange this for them, so must remember to arrange cardiac appointments themselves. Have you considered this?

This is an important issue. Female carriers are not always routinely followed up. This can be due to a range of factors including: misinformation suggesting that further checks are not necessary because initial test results are normal; so much going on in the family that it's difficult for mums to look after themselves; NHS systems don't issue follow up appointments or don't have a system to allow appointments to be made three years ahead.

It is important that the reason and rationale for testing and follow up is explained to carriers, empowering them to request appointments, if they are not being arranged to schedule. However, making unnecessarily frequent appointments can over-burden families and even increase 'failure to attend' rates. Remember that most carrier-women will not develop a cardiomyopathy, but it is very important not to miss those who will and pick up early deterioration in cardiac function.

Health Care Professionals as well as carriers need to understand that cardiac follow up needs to be continued to schedule, even in those whose latest heart test results are normal.

Question 3: Not all cardiologists are following the Standards of Care at the moment – are the guidelines from DMD Care UK being widely disseminated to cardiologists?

Yes, they are being disseminated progressively. It is true that currently, practice differs between cardiologists. This is partly because they are not used to providing ongoing surveillance and deploying preventative treatments for other cardiac conditions. In DMD, we know exactly why the heart problem (cardiomyopathy) occurs and what can be done to reduce and slow its rate of progression. For therapies to be most effective, they need to be initiated at the earliest signs of

that the heart is becoming affected and long before patients would be aware of any symptoms or limitations.

We have published these management recommendations in an open-access journal to ensure that they are accessible and widely available to families as well as to cardiologists and other health care professionals. The rationale and evidence on which the recommendations are based are also referenced for those who need that background information.

The guideline paper has been endorsed by the *British Cardiovascular Society* and will appear on their website. Furthermore, cardiologists working alongside *North Star Centres* have been made aware of this cardiac guidance.

Question 4: When should routine Electrocardiogram (ECG) be started?

Ideally, as a routine test at the first cardiac assessment (ie: at or before age 6 years). Along with the other information contained on the ECG, it also provides a definitive measure of heart rate at each assessment. This allows detection of progressively increasing heart rate trend over time which occurs commonly in boys with DMD. The resting heart rate determines whether the dose of beta-blocker dose needs to be increased to achieve the target resting heart rate of 75-85 beats / minute.

Question 5: I have come across patients with severe cardiomyopathy, on the three recommended cardiac medications, who are given oral potassium following diarrhoea and not monitored afterwards – should this be written into their Emergency Healthcare Plans?

The use of oral potassium supplementation is rarely indicated and is probably derived from a time before potassium sparing drugs became available. Oral potassium is unpleasant to take and, more importantly, is poorly absorbed. It's much more efficient, therefore, to use potassium sparing diuretics (eg: spironolactone or eplerenone) to replace or maintain serum potassium levels.

Question 6: How useful are heart-failure medications and implantable cardioverter defibrillators (ICDs) in adult populations with advanced DMD?

In the paediatric guidance, we also include a second table to indicate options for patients *at any age* with advanced left ventricular dysfunction due to dystrophinopathy. It summarises the place of some newer, more potent, heart failure treatments [eg: sacubitril-valsartan (*Entresto*); sodium-glucose cotransporter-2 inhibitors (*Gliflozins*)], indications for the potent anti-arrhythmic drug, amiodarone, and implantable device therapy (ie: cardiac resynchronisation pacing for heart failure; implantable cardioverter-defibrillators). Please consult that flow-diagram for its additional guidance in paper.

https://openheart.bmj.com/content/9/2/e001977.long