

The Inherited Cardiovascular Diseases Service

Information for families

Great Ormond Street Hospital
for Children NHS Foundation Trust

This leaflet explains about the Centre for Inherited Cardiovascular Disease at Great Ormond Street Hospital (GOSH) and what to expect when you and your child come to see us.

Our aim is to develop a close and supportive partnership with parents and their children to ensure a positive experience for both the families/carers and ourselves. We recognise that the hospital environment can prove to be demanding, worrying and intense and we aim to alleviate some of the stress to families by talking openly, listening to each other and working together in the best interests of the child. This way we hope to create a partnership with families that is built on mutual respect and trust.

Who are we?

The Inherited Cardiovascular Disease Service is a specialised team involved in the care of your child from their first assessment at GOSH.

The team is made up of:

- **cardiologists**, who are doctors specialising in the diagnosis and non-surgical treatment of inherited cardiovascular disease
- **electrophysiologists**, who are doctors specialising in diagnosis and treatment of arrhythmias using medicine or surgery
- **clinical nurse specialists**, who are experienced cardiac nurses specialising in inherited cardiovascular conditions
- **clinical psychologists**, who help children, young people and families to come to terms with the diagnosis and any potential impact it may have on day to day living
- **genetic counsellors**, who explain about genetic conditions, how they can be passed on and how they could affect your family
- **cardiac technicians**, who carry out the screening tests, such as ECHOs and exercise testing
- **administrative support staff**, who organise clinic appointments, maintain medical records and type up reports from the other members of the team.

We specialise in the following conditions:

Cardiomyopathy

- Hypertrophic cardiomyopathy (HCM/HOCM)
- Dilated cardiomyopathy (DCM)
- Arrhythmogenic right ventricular cardiomyopathy (ARVC)
- Restrictive cardiomyopathy (RCM)
- Left ventricular non-compaction (LVNC)
- Other/non-specific cardiomyopathies

Inherited arrhythmia syndromes

- Long QT syndrome (LQTS)
- Brugada syndrome
- Catecholaminergic polymorphic ventricular tachycardia (CPVT)
- Short QT syndrome (SQTS)
- J-wave syndromes (early repolarisation syndrome)
- Family history of unexplained sudden cardiac death (SCD/SADS)

Aortopathy/connective tissue disorders

- Marfan syndrome
- Loeys-Dietz syndrome
- Ehlers-Danlos syndrome
- Arterial tortuosity syndrome
- Familial supravalvular aortic stenosis
- Beales syndrome
- Other aortopathies

How do I get my children screened?

We need a referral from your general practitioner (GP), paediatrician or local cardiologists for your children to be screened in one of our clinics

Before your appointment

One of the specialist nurses will try to contact you by telephone to answer your questions and to explain what will happen during the clinic appointment. We may ask you some questions about the history of heart problems in your family. It will be useful to obtain clinical information relating to any members of the family who already have a cardiac diagnosis, and we may need to ask for your permission to access this information. You are welcome to call the clinical nurse specialists before your appointment if you have any questions or concerns. You will find our contact details at the back of this leaflet.

The appointment

Who will we meet?

The clinical nurse specialists will meet you at the start of your day and will be available throughout the day, to make your clinic visit as easy as possible. They are experienced cardiac nurses who will be your contact if you have any concerns or questions after you have gone home. They will be present in follow up clinics and for any stays in hospital.

You will also meet an experienced team of technicians who will carry out the tests for you. They will explain what they are doing and answer any of your questions.

What shall I tell my child about the appointment?

The medical, nursing and technical staff are specially trained to work with children will always tell you and your child what to expect before they start any tests. Tell your child as much as you can about what will happen, using simple words your child can understand, and answer any questions truthfully. If your child is particularly anxious about a test, the clinical nurse specialists are happy to speak to them before the test and help during the tests. Please let us know in advance if you think this would be of benefit to your and your child.

For more information about the hospital and staff, visit www.gosh.nhs.uk. We also have information for children and teenagers available on the website.

What cardiac screening tests will my child have?

The cardiac screening tests are to assess if your child has the condition and also provides us with measurements of improvement, stability or deterioration for children who have the condition. An ECG will be recorded at every visit. Other scans or tests may be requested – your nurse can tell you which will be done at each appointment. You are able to stay with your child during all the tests.

- An **ECG** records the electrical signal as it is conducted throughout the heart. It is a simple test performed by placing sticky electrodes on the child's chest, legs and wrists. An ECG is entirely safe, takes a few minutes and causes no pain, although the child may be anxious about the stickers and connecting wires.
- An **Echo** is an ultrasound scan of the heart. A picture of the heart is produced from which an accurate assessment of the size and function of the heart can be made. The scan takes from 30 to 40 minutes. As before this test is not painful, but children sometimes find the jelly on the probe a bit uncomfortable. Children can watch their favourite videos during the scan.
- An **MRI** scan uses a magnetic field rather than x-rays to take pictures of your child's body. The MRI scanner is a hollow machine with a tube running horizontally through its middle. Your child will lie on a bed that slides into the tube. An MRI scan usually lasts between 20 minutes and an hour. Please watch our podcast (short video film) about having an MRI, available at www.gosh.nhs.uk/medical-conditions/procedures-and-treatments/your-child-is-having-an-mri-scan/video-your-child-is-having-an-mri-scan/ or on the GOSH channel on YouTube. The MRI machine is very noisy, but your child will be given headphones and can watch a film of their choice while the scan is carried out. A cardiac MRI scan can be carried out in around 15 minutes, but some children may require a longer, more detailed scan. This type of scan is usually only carried out in older children, because they need to stay very still while the scan is taking place. If very young children need to have an MRI scan, this may need to be done under a general anaesthetic as they may find it difficult to stay still for such a long time.
- A **24-hour ECG** recording (or Holter) uses a small box similar in size to a portable stereo, from which three leads are attached by sticky pads to your child's chest. This is an ECG monitor that continuously records the heartbeat over 24 to 48 hours. Children wear the monitor under their clothes and can continue with their normal daily life including sport and exercise (although not swimming). You and your child will be asked to document your child's activities during the 24 to

48 hour period. This is very important as we will match this with the recording when the box is analysed. When the test is finished, you will be required to return the monitor to GOSH – please do this promptly so another child can use the monitor.

- An **exercise test** assesses the rhythm and function of the heart during exercise on a bicycle or treadmill. Blood pressure and breathing are also monitored during the test. This test is usually only performed on children over the age of eight years due to their size. Exercise testing also provides us with an objective measurement of improvement, stability or worsening of heart function over time. The test takes approximately 45 minutes and allows symptoms not obvious at rest to become apparent when the heart is working harder. We recommend children to wear loose comfortable clothing for the test.

When will we be given the results of the tests?

After your child's cardiac screening tests are completed you will see the consultant and clinical nurse specialist at your consultation. The results will be given and discussed with you. We will also send a letter to you and to your child's GP. At the end of the consultation the team will discuss with you when they want to see your child again and if so, approximately when.

Most conditions require regular life-long monitoring and some may need treatment with medicines or surgery or both. In many cases, children will have had normal screening test results, but where there is a known family history of inherited heart problems, we will continue to monitor them on a regular basis as they grow older.

We hope you will feel able to ask questions of any member of the team about anything you do not understand. It is important to ask questions and to make sure you get explanations in language that you understand. When you see the team in clinic you may find it useful to take in a list of questions you want to ask.

How long will we be at the hospital?

As far as possible, we try to group tests and clinic appointments on the same day to minimise the number of times that families have to visit the hospital. This does mean that you will be with us for at least two to three hours on your appointment day. It can take slightly longer than this if your child is undergoing additional tests. The department often gets very busy and you may have to wait to be seen at times. Please be patient and bear with us! There are play areas within the hospital and department. Children may also like to bring along their favourite books or toy.

Where can we eat?

The Lagoon Restaurant on Level 2 (Ground Floor) of the Morgan Stanley Clinical Building (MSCB) serves a range of snacks and full meals. You can also bring your own snack or lunch if you like. If there is a long queue for tests or to see the doctor, there may be time for you to go to get a drink or some food. The staff in the clinic will be able to tell you if the wait is likely to be long enough to allow for this.

Frequently asked questions

How do I advise family members about screening?

When a child or adult is diagnosed with Inherited Cardiovascular Disease it is advised that their family members be assessed for the same condition. Each family member will have an ECG and ECHO as part of their assessment. Each family has our contact details that can be given to other family members so they can contact us for advice.

How often do we screen children?

We screen children from birth. Thereafter we screen children and young people at regular intervals. Screening may continue into adulthood, although not so often. The team will discuss the frequency of your child's screening with you at the time of your clinic appointment.

When children reach the age of 16 to 18 years, their care will be transferred to an adult service. Most children are transferred to the care of Bart's Hospital, around a mile from GOSH, where there is a large service specialising in inherited cardiovascular disease. In some cases we transfer care to other hospitals, particularly where other family members are already looked after by a particular team. The clinical nurse specialists are closely involved in the transition of your child's care from one team to another.

What if my child develops worrying symptoms during screening intervals?

If you are worried about your child please contact the team who will advise you and decide if it is necessary to bring your appointment forward or arrange further tests.

We are very happy to reschedule appointments wherever necessary. We always try to see siblings in the same clinic to minimise inconvenience, so please contact our administration team if you would like us coordinate appointments for more than one child.

What do I tell my child's GP/local healthcare team or school about my child being screened or diagnosis?

The team will send you and your GP and local teams a copy of your clinic letter. The clinical nurse specialists are also happy to liaise with local healthcare teams and nurseries/schools and sports clubs providing further information and support. We work closely with the Cardiomyopathy Association, SADS UK, CRY and other support agencies.

What genetic screening is available for children and families?

Genetic testing may be available in some cases so please discuss this with us. We work closely with genetic specialists and if a family decides to have genetic screening we liaise and work closely with a genetic counsellor to discuss the implications of this type of test.

Genetic testing can be very complex. In some cases, individuals who carry a gene change will not go on to develop the condition and may never have any symptoms or features of the disease. There are many implications to consider before having such a test and the team will support you through these decisions.

Can you help with my child's anxieties and worries?

Many of the families visiting our clinics are dealing with very difficult situations, including bereavements, new diagnoses and worrying symptoms. It is very natural for our patients, their parents and siblings to feel anxious and concerned about these issues. Our team includes specialist clinical psychologists who are able to offer tailored support and advice to children and adults affected by inherited cardiac conditions. The psychologists can talk to patients/parents over the phone, meet with them at our cardiac clinic appointments or can arrange separate counselling appointments in their own department. The psychology

service is routinely offered to all families attending our clinics. Please let the nurse specialists know if you think that this is something of interest.

Are there any opportunities for my child to be involved in research?

Our team looks after a unique group of patients, many of whom are diagnosed with conditions which are rarely seen in children in the wider population. Due to the unique nature of our service, we are well-placed to carry out research projects which aim to help advance our understanding of inherited cardiac conditions and to explore new treatments and screening techniques. From time to time, members of our specialist research team may approach families in the clinic to see whether they are willing to take part in relevant research projects. The decision to take part in research projects is entirely voluntary and the treatment of our patients will in no way be affected if they choose not to participate.

Further information and support

If you have any questions or would like some more information, please contact the Clinical Nurse Specialists at Great Ormond Street Hospital on 020 7405 9200 ext. 5124, 5305 or 5139. You can also email the service at icvd@gosh.nhs.uk

The Cardiomyopathy Association

supports people with all types of cardiomyopathy. Telephone their helpline on 0800 018 1024 or visit their website at www.cardiomyopathy.org

CRY – Cardiac Risk in the Young offer support and advice to families affected by young sudden cardiac death. Telephone them on 01737 636 222 or visit their website at www.c-r-y.org.uk

SADS UK offers support and advice about heart conditions that can lead to sudden unexpected death. Telephone them on 01277 811 215 or visit their website at www.sadsuk.org

The British Heart Foundation provides information, advice and support to anyone affected by heart disease. Call their helpline on 0300 330 3311 or visit their website at www.bhf.org.uk

The Marfan Association UK supports anyone affected by Marfan Syndrome. Call them on 01252 810472 or visit their website at www.marfan-association.org.uk

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Notes

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