The Inherited Cardiovascular Diseases Service

Information for families

Great Ormond Street Hospital
for Children NHS Foundation Trust

This leaflet explains about the Inherited Cardiovascular Disease Service 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 children's nurses specialising in cardiac nursing
- **clinical psychologists**, who help children, young people and families to come to terms with the diagnosis and its potential impact
- **genetic counsellors**, who explain about genetic diseases 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
- Dilated cardiomyopathy
- Arrhythmogenic right ventricular cardiomyopathy
- Restrictive cardiomyopathy
- Left ventricular non-compaction
- Other/non-specific cardiomyopathies

Inherited arrhythmia syndromes

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

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 (specialist children's doctor) or local cardiologists for your children to be screened. Some of the team are also based at the Heart Hospital, which specialises in Inherited Cardiovascular Disease in adults.

When children reach the age of 16 to 18 years, their care will be transferred to an adult service, either to the Heart Hospital or their local cardiology team. The clinical nurse specialists are closely involved in the transition of your child's care from one team to another.

Before your appointment

You are welcome to call the clinical nurse specialists before your appointment to discuss what will happen and pass on any information about your family's health.

Frequently asked questions

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 children's cardiac nurses and 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?

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 of the tests. Tell your child as much as you can about what will happen, use 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 using distraction techniques during the tests. 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. You are able to stay with your child during all the tests.

Your child will have an Electrocardiogram (ECG) to look at the electrical signal of the heart, and an Echocardiogram (Echo), an ultrasound scan of the heart. They may also require an exercise test to assess the rhythm and function of their heart at a faster rate. We may also need to perform additional investigations, which may include blood tests, having a 24-hour ECG monitoring (Holter), and a magnetic resonance imaging (MRI) scan of the heart.

- 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-childis-having-an-mri-scan/video-your-childis-having-an-mri-scan/ or on the GOSH channel on YouTube.
- 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. You and your child will be asked to document your child's activities during the 24 to 48 hour period to match with the recording when the box is analysed. When the test is finished, you will be required to return the monitor to GOSH so that the results can be analysed.

- An **exercise test** is a specially modified test that assesses the rhythm and function of the heart at a faster rate, while the child is on an exercise bicycle or a 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.
- **Electrophysiological studies** are a special type of cardiac catheterisation used to study the electrical activity of the heart in detail. It is needed when a potentially more serious abnormal heartbeat is suspected. This procedure will involve your child having a general anaesthetic.
- A **tilt test** helps to understand fainting spells and involves lying on a table which is then tilted into various positions.
- An **Ajmaline test** is very similar to an ECG except that your child is given an injection of a medicine called Ajmaline, which brings about changes on the ECG that can help us to diagnose a specific condition called Brugada syndrome. The medicine is given through a cannula (thin plastic tube) during which your child's heart rate is closely monitored for changes over a period of 15 minutes.

When will we be given the results of the tests?

After your 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 send a copy of the clinic letter to you and 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. 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.

What happens next?

Once your child has had all the screening tests, the team will look at the results to try to reach a diagnosis. They will explain the diagnosis and what happens next. Most conditions will require regular life-long monitoring and some may need treatment with medicines or surgery or both.

How long will we be at the hospital?

You will be with us for at least two to three hours on your appointment day. The department often gets very busy and you may have to wait longer. 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 lunch if you like.

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.

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.

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 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?

We are still learning about the disease process and inheritance pattern. 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 a genetic test.

We know that not all those who carry the gene will develop the

condition. This means that some people may have no symptoms or even no features of the disease but may still carry the abnormal gene(s) that determine the disease. There are many implications to consider before having such a test and the genetic counsellors and other members of the team will support you through these decisions.

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. 5646 or 5305. 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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