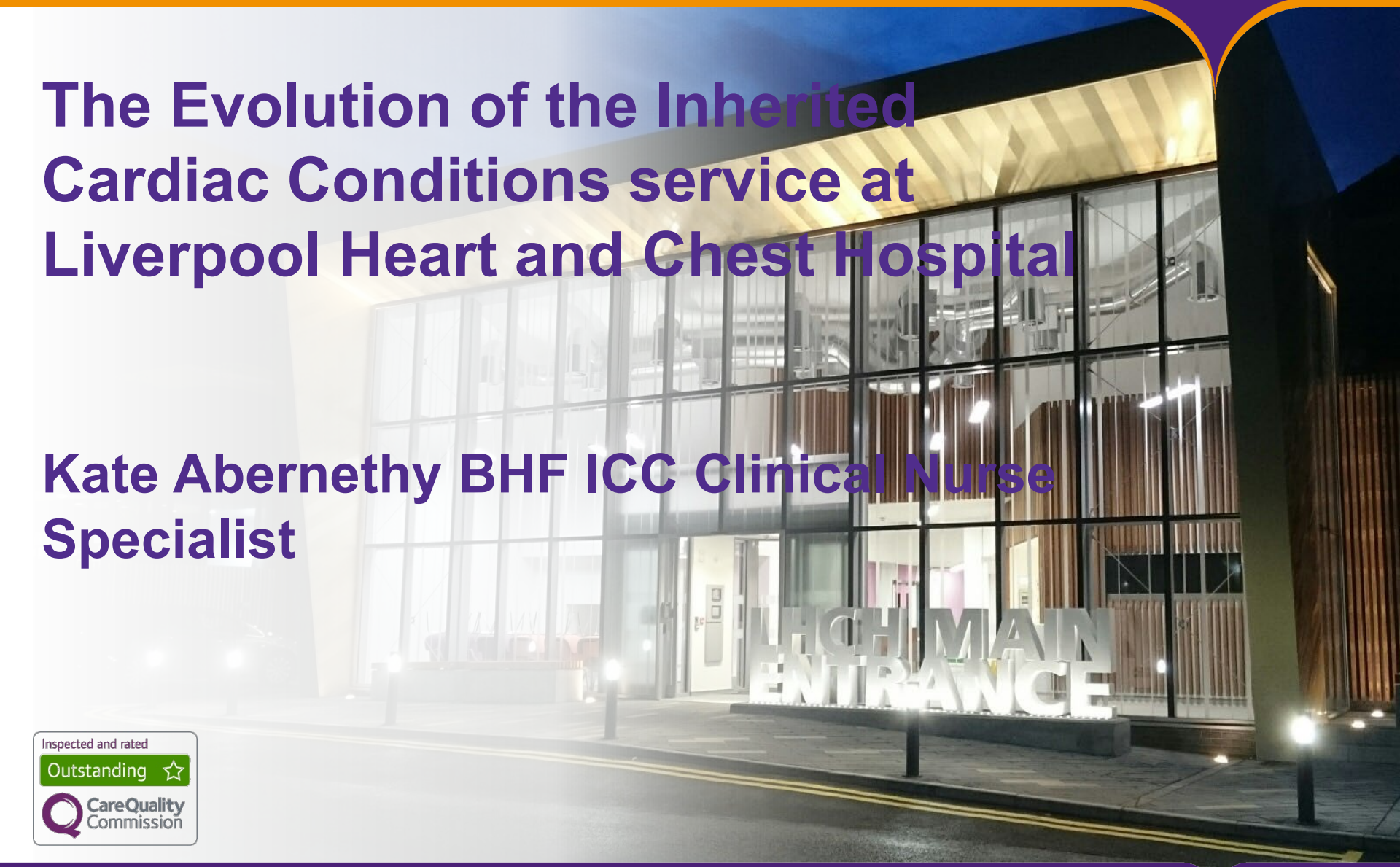
The Evolution of the Inherited Cardiac Conditions service at Liverpool Heart and Chest Hospital

Kate Abernethy BHF ICC Clinical Nurse Specialist



Inspected and rated Outstanding
CareQuality Commission

What we do

Liverpool Heart and Chest Hospital

OUR VISION IS 'TO BE THE BEST'

Leading and delivering outstanding heart and chest care and research

WE SERVE A CATCHMENT AREA OF 2.8 MILLION PEOPLE

ACROSS MERSEYSIDE, CHESHIRE, N. WALES & THE ISLE OF MAN... & BEYOND FOR OTHER SPECIALIST SERVICES

EACH YEAR WE SEE...

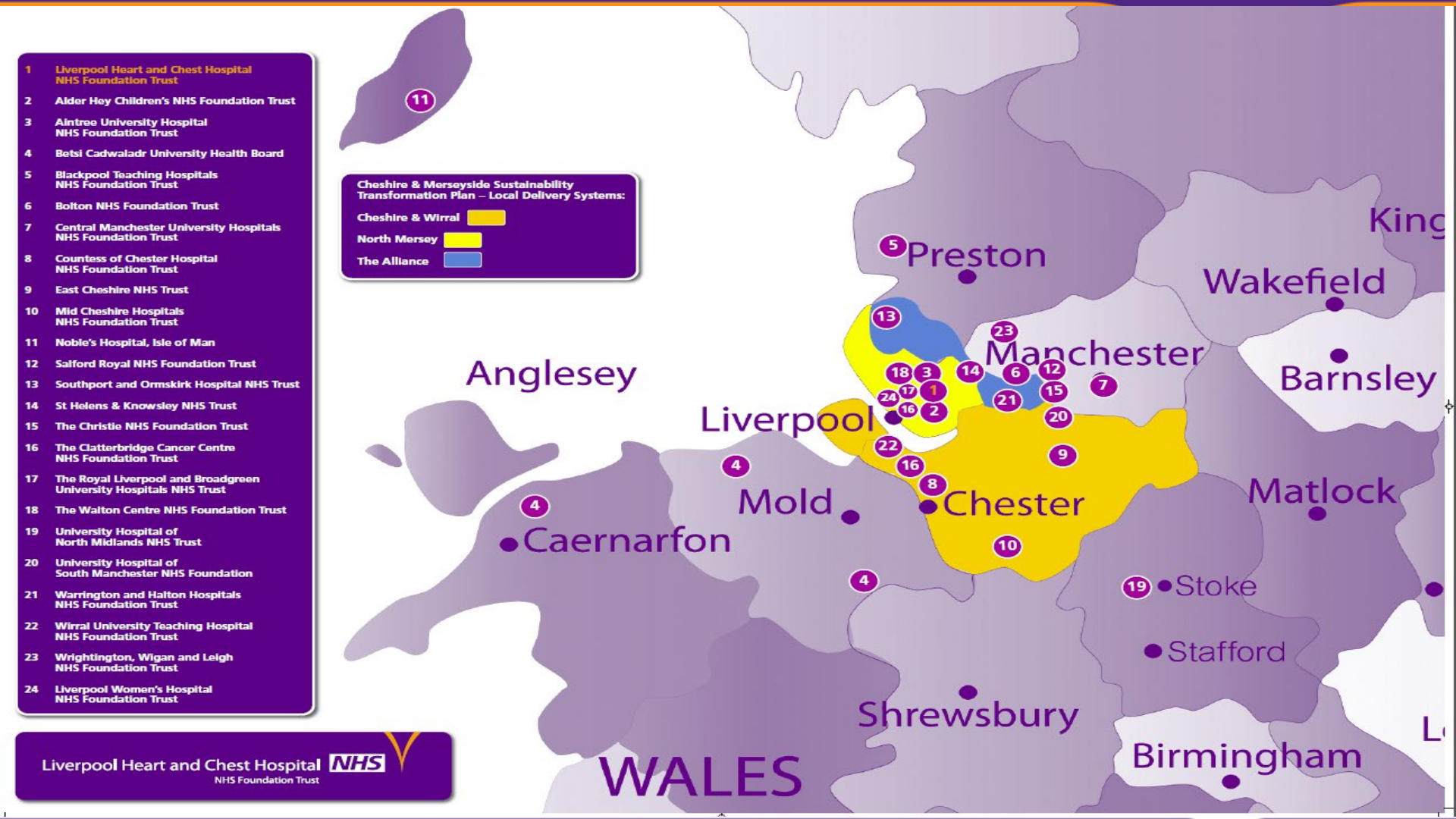
- 100,000 OUTPATIENTS
- 13,000 INPATIENTS

Again in 2019

IN SEPTEMBER 2016, LHCH WAS RATED OUTSTANDING BY THE CQC

13 (+4) Referring Trusts, Serving 2.8 Million People

Liverpool Heart and Chest Hospital NHS
NHS Foundation Trust



Liverpool Heart and Chest Hospital NHS
NHS Foundation Trust

Excellent, Compassionate and Safe Care for every patient, every day

Liverpool Heart and Chest Hospital NHS

AN OUTSTANDING TRUST IN BRIEF

2,190 cardiac surgery operations

1,260 thoracic surgery operations

320 cystic fibrosis patients

2,600 angioplasty

1,220 catheter

185 aortic aneurysm procedures

7,220 MRI, CT, Echo Scans

68,918 hospital outpatients

8,958 cardiology inpatients

193 inpatient beds

9 operating theatres

5 catheter laboratories

10 community locations

1,599 staff are employed

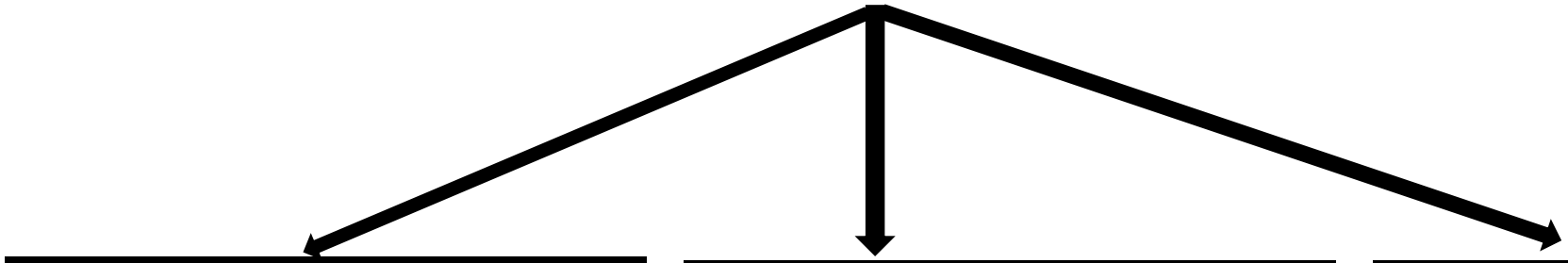
£129m turnover

2.8million population served by LHCH

Largest single-site heart and chest hospital

Excellent, Compassionate and Safe Care for every patient, every day

Inherited cardiac conditions are a group of genetic disorders that primarily affect the heart



Cardiomyopathies

Hypertrophic Cardiomyopathy
Dilated Cardiomyopathy
Arrhythmogenic Cardiomyopathy
Restrictive Cardiomyopathy

Channelopathies

Long QT syndrome
Brugada syndrome
CPVT
Early repolarisation syndrome

Aortopathies

Marfan syndrome
Loey-Dietz syndrome

AICC Mission Statement

To provide consistent, top quality education and training, advice on management and best practice, as well as acting as a forum for data collection, audit and collaborative research.
Membership is open to clinicians, nurses, counsellors, scientists and professionals allied to medicine, as well as persons from organisations and charities involved in support of such families.

Association for Inherited Cardiac Conditions

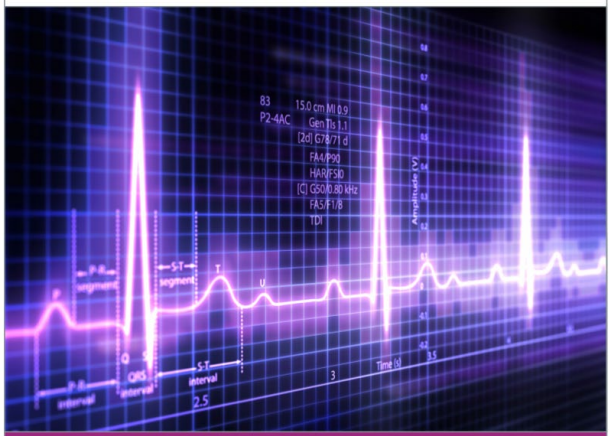
Acknowledging the problem

phg foundation
making science work for health

Heart to Heart

Inherited Cardiovascular Conditions Services

A Needs Assessment and Service Review

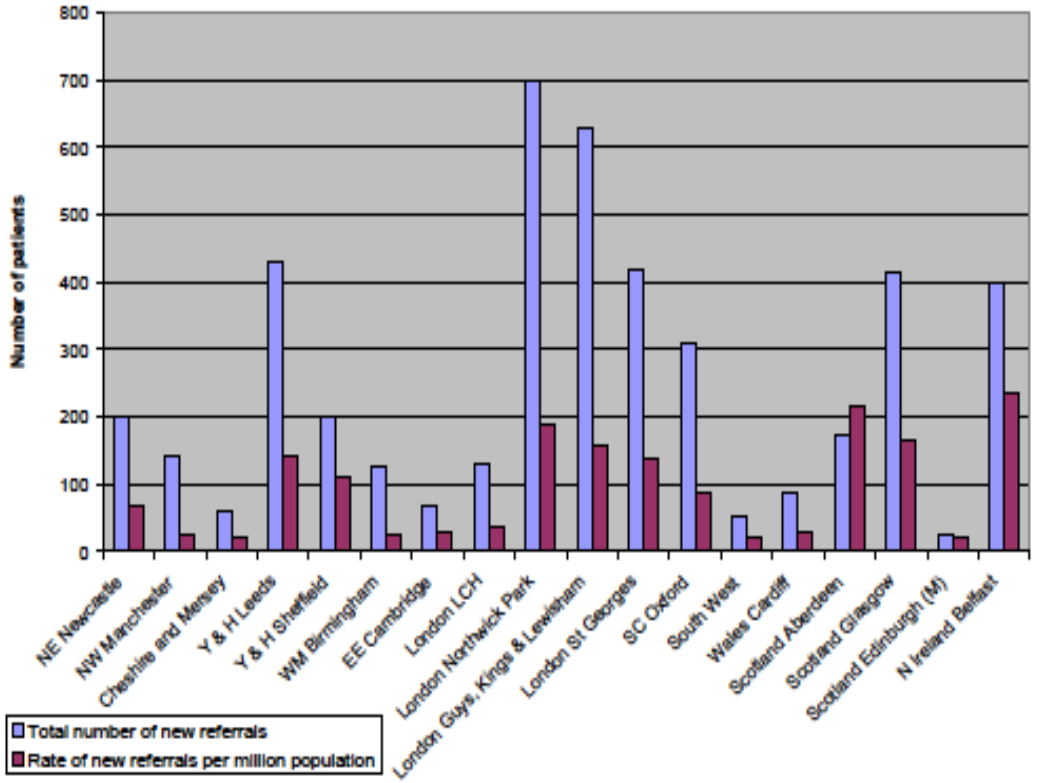


Hilary Burton
Corinna Alberg
Alison Stewart

June 2009

www.phgfoundation.org

Figure 5.3 Annual estimated number of new referrals and rate per million catchment population



The numbers from THH have not be included due to the national nature of this service - see paragraph 5.4.1 on The Heart Hospital

British Heart foundation Cardiac Genetics Nurses.

- In 2008 the BHF launched an initiative to enhance the care and services delivered to patients and their families with or at risk of an ICC.
- They funded 9 BHF Cardiac Genetics Nurse posts, across England & Wales; previously funding 2 CGN posts in Leeds & Belfast – 2006, for a 3 year period.

- Evaluation of the BHF CGN Service Development was undertaken and published in 2012 – the Project team involved:
  Prof Maggie Kirk & Dr Emma Tonkin.

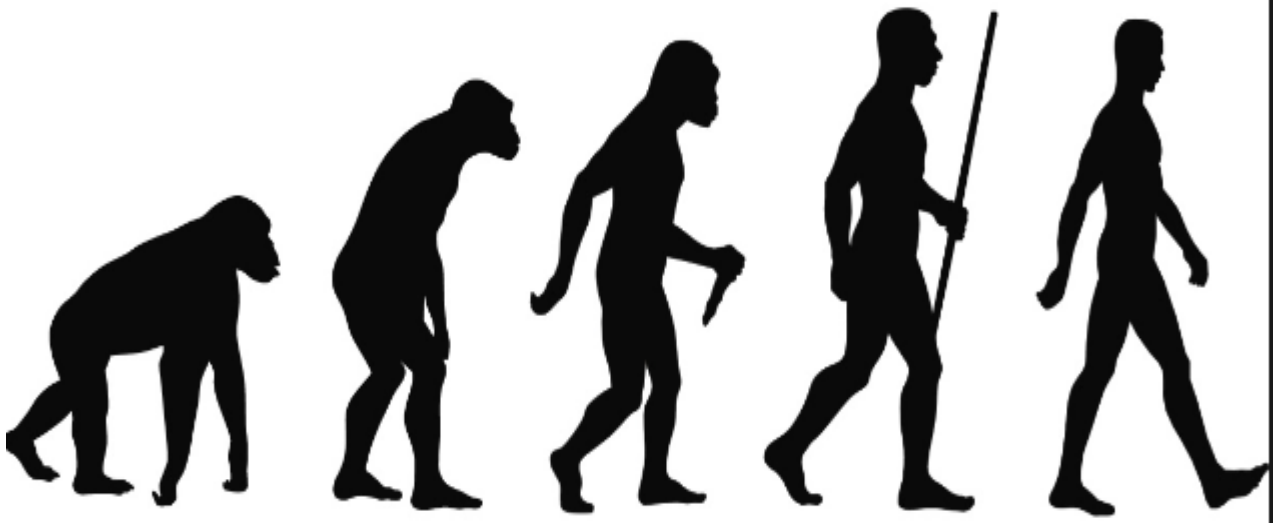
- It found that the CGN model was fit for purpose & that they make an effective, value for money contribution to the ICC service

- Providing efficient service delivery across 2 specialisms.–
  (Cardiology & Genetics).

British Heart Foundation

Evolution of the ICC Service At LHCH

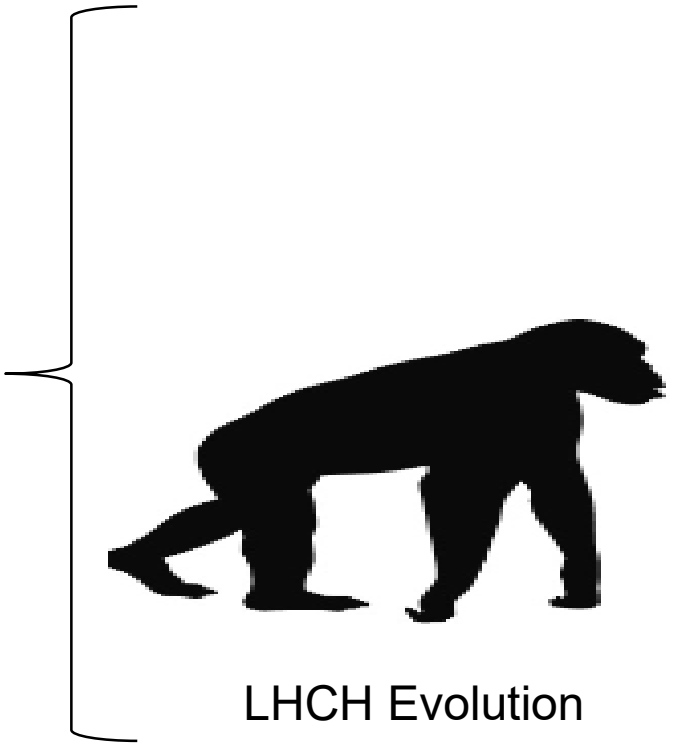
2009 → Present



- Single monthly named ICC clinic
  - One EP Cardiologist
  - One EP Nurse
- Lots of HCM managed in general clinics
- Genetics services available
  - Distant
  - No MDT
  - Most Cardiologists were unaware

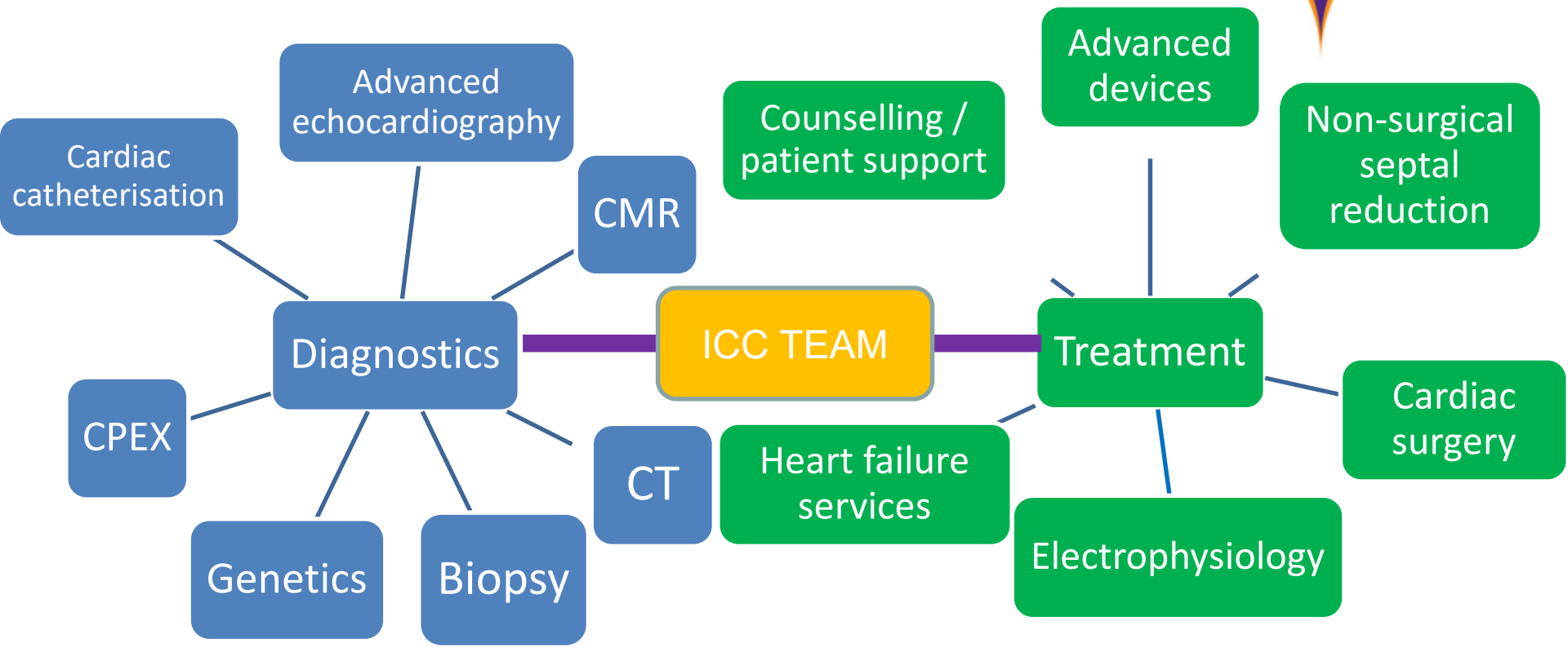
- Good capacity for clinical management, but disorganised care

- There were tools available within our grasp but nobody knew how to use them

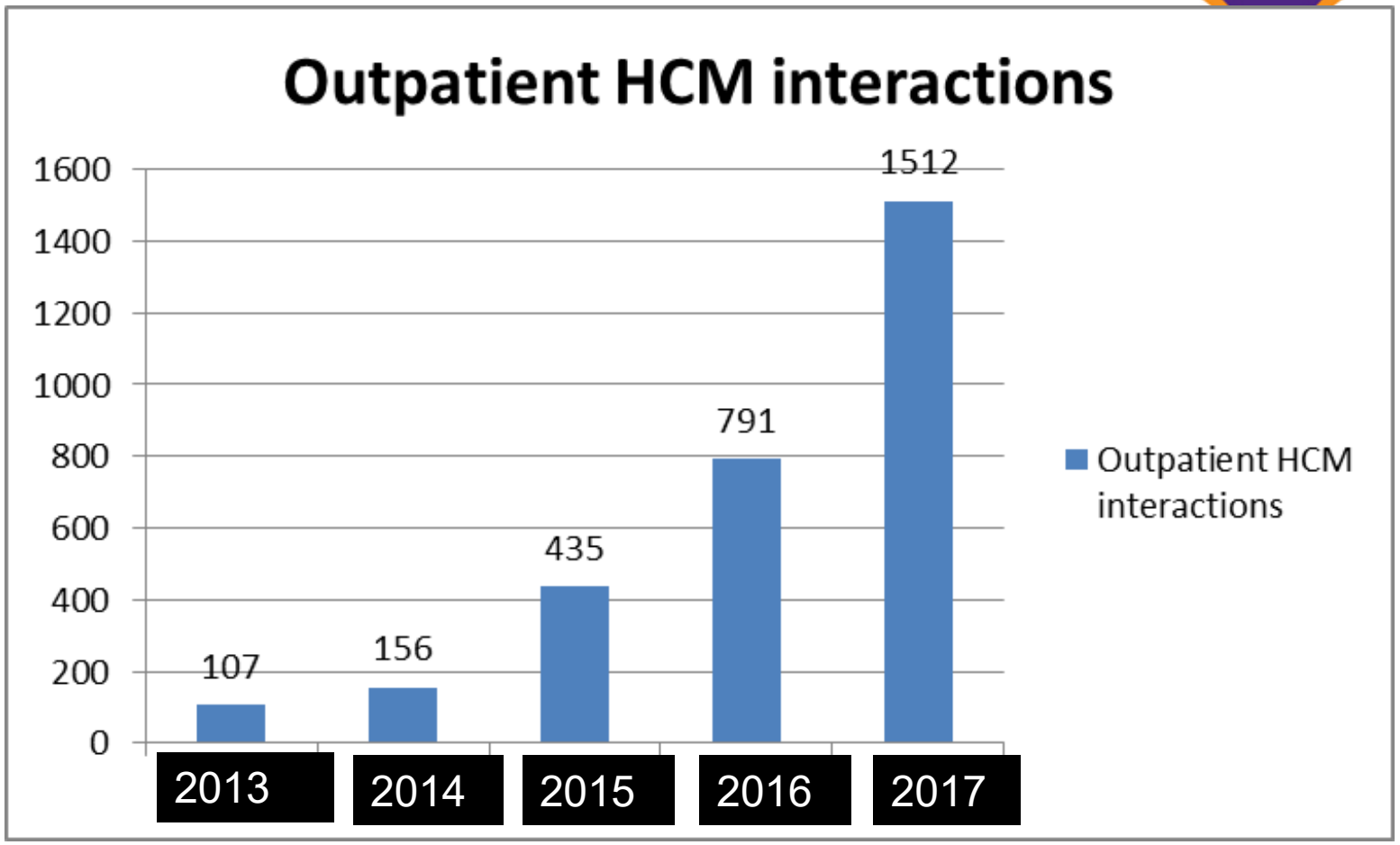


LHCH Evolution

Components of an ICC Service

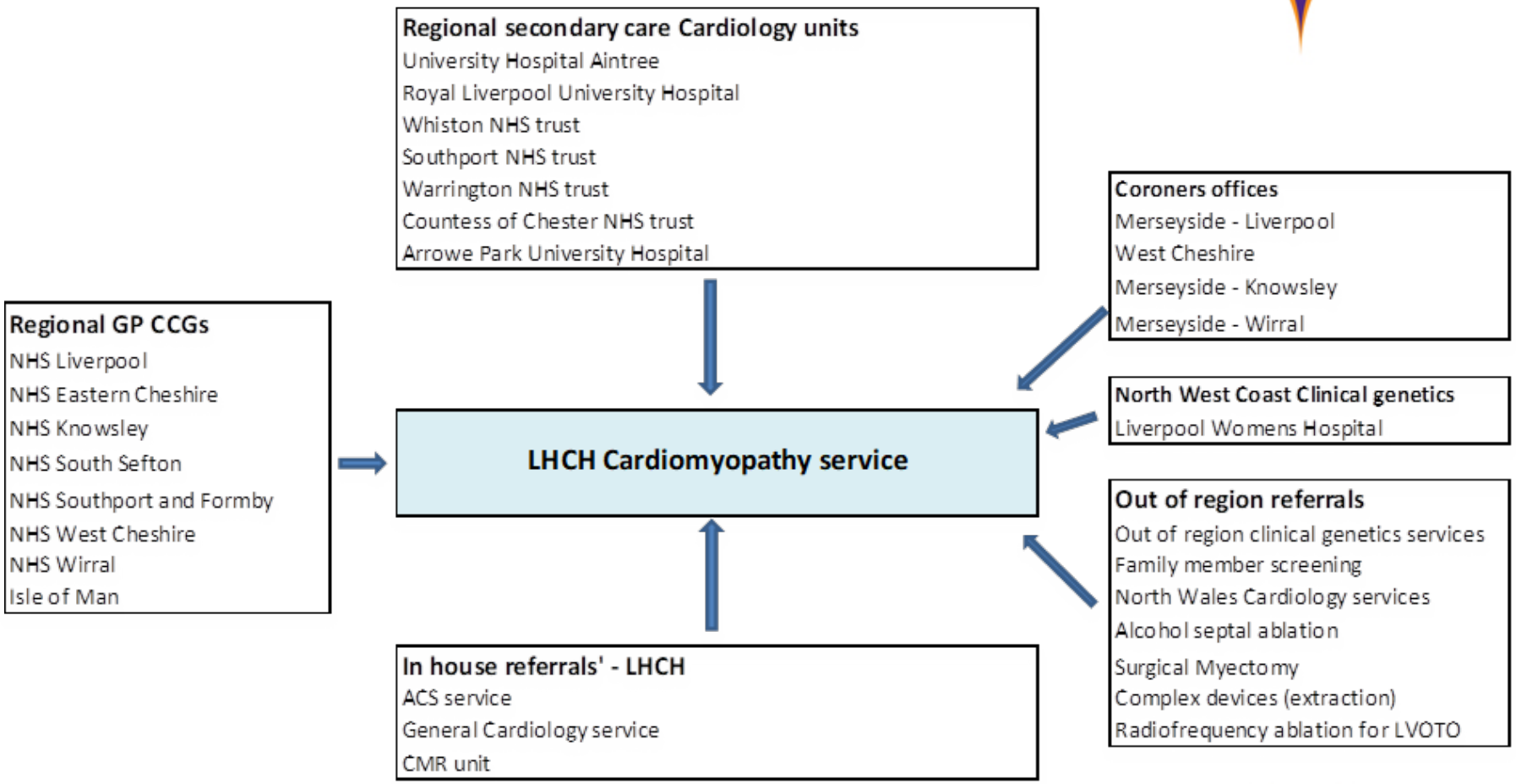


Patient Group is rapidly expanding



LHCH source of Referrals

Source of referrals to LHCH Cardiomyopathy unit



Improving Referral rates

LHCH is the only site in Cheshire and Merseyside to perform diagnostic cardiac genetics

Part of my role is to raise awareness:

- Both clinical management and genetic screening
- ICC afforded the same time as ACS / HF / Devices etc
- Annual ICC study day – Since 2014.
- Multiple talks to GP groups in different areas
- Multiple talks to nurse specs
- Talks for Charities – CMUK, BHF – for HCPs & public
- Meetings with coroners
- Arranging MDTs at Alder Hey Children's hospital
- Transition clinic

- https://extras.bhf.org.uk/milesstory/
- The Miles Frost Fund, in partnership with the British Heart Foundation, will create a life saving legacy.
- They aim to raise £1.5 million to fund nurses and genetic counsellors across the country, so that more families at risk of hypertrophic cardiomyopathy get the testing and treatment they need.

Improving our ICC service

Implement a transition from paediatric to adult service between Alder Hey Children's Hospital & LHCH, was identified as gap in service provision.

Improve cohesion within Cardiology

- Bring all of the pre-existing tools together

Integration with Clinical Genetics Team

- Twice weekly onsite LHCH Clinical geneticist & Genetic Counsellor present
- Increasing awareness in both Cardiology and Genetics
- Adaptation in MDTs
  - Fortnightly at LHCH
  - Planned:
  - Bimonthly at Alder Hey Children's Hospital and Liverpool Women's Hospital (LWH)

Transition

- What the government bodies say…
- CQC: ‘From the pond into the sea’ 2014; NICE & NHS England:

- Found a shortfall between policy & practice.
- Only 50% of young people & their parents said they had received support from a lead professional during the process leading up to transition to adult services.
- Commissioners must listen & learn from young people & their families
- Existing good practice guidance must be followed to ensure young people are properly supported through transition
- Adolescence should be recognised as an important development phase across the health service.

We want to be more Accessible to patients

Communication

- Time and opportunity – large part of my role

Strong links to local CMUK patient support group

- CMUK representatives at larger CMP clinics at LHCH
- Venue for support group
- Cardiologist / Geneticist / nurses / clinical psychologist / Research - all contribute annually since 2017

Social media

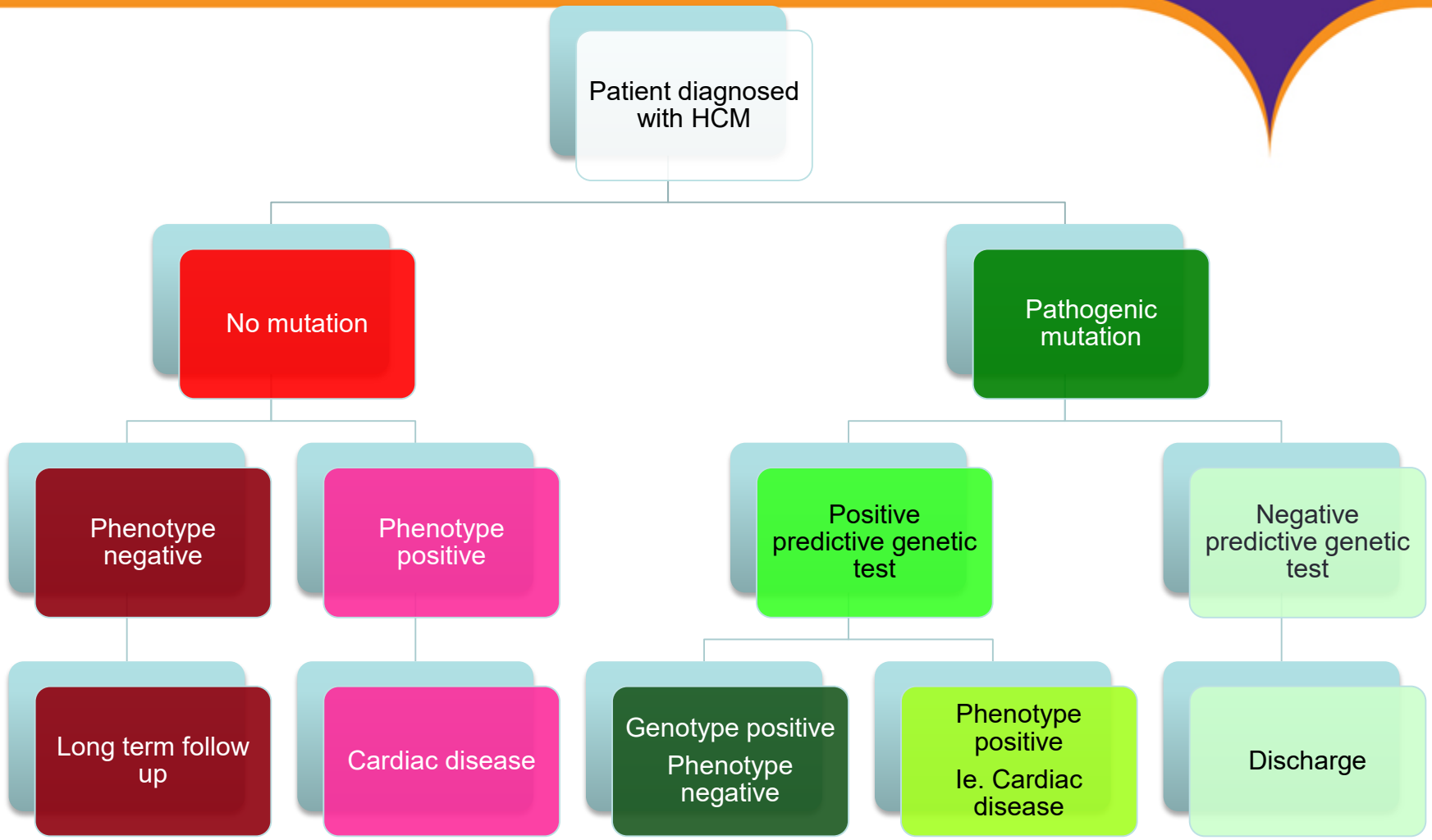
- LHCH trust twitter account is active
- Propose an ICC twitter account – trust have already given permission for this – on agenda

LHCH ICC website updated

- http://www.lhch.nhs.uk/our-services/cardiology/inherited-cardiac-conditions/

LHCH ICC phone line and co-ordinator

Genetic screening process



LHCH – previous approach to genetic testing

Process	Possible loss
Informative genetic test	
↓	→ Cardiologist may not process result
Proband referred to clinical genetics	
Letter in the post	
↓	→ Cardiologist may not refer proband
	→ Proband may not accept referral - possible confusion
	→ Proband may opt out
Proband seen in clinical genetics	
Pedigree	→ Dear family member letter may not be received
Dear family member' letters	→ Confusion with GP referral
↓	Relatives may opt out - possible confusion
Relative seen in clinical genetics	
Genetic counselling	
Predictive testing	

HCM Diagnostic Genetic Testing Audit

Unsolicited and presented internally.

All diagnostic testing performed at LHCH 2013-16

Results and progression to predictive testing.

101 tests; 31 informative

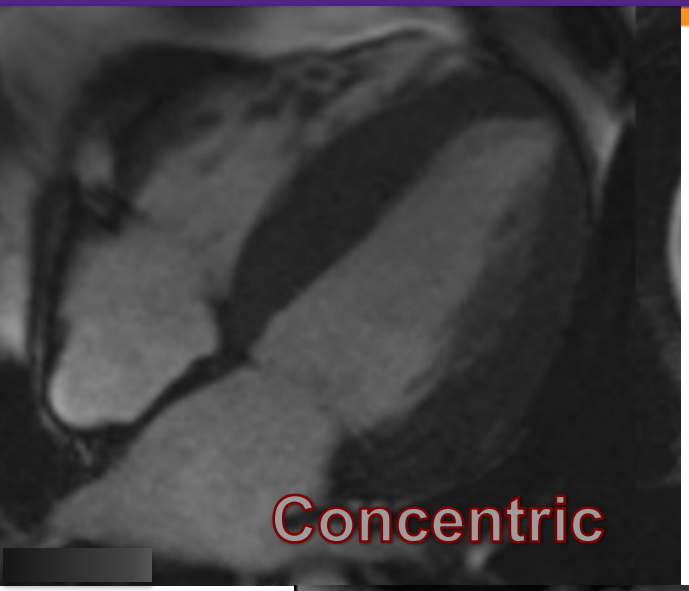
<u>Only 9/31 informative diagnostic test results reviewed in clinical genetics for predictive testing</u>

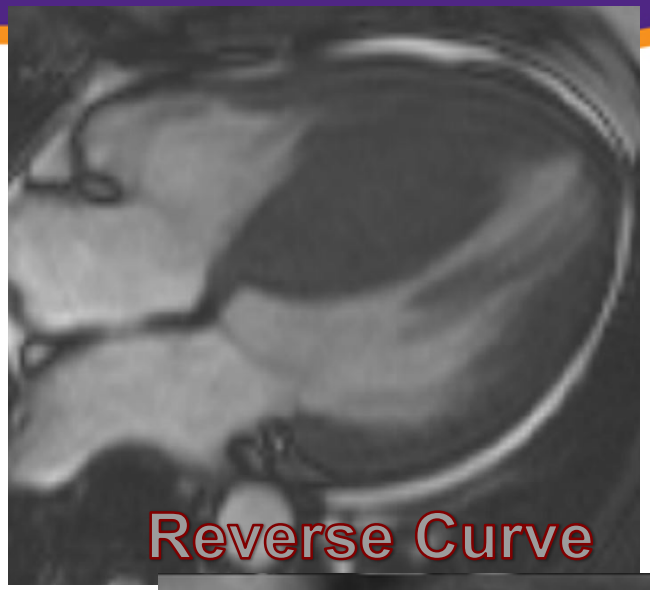
21 relatives tested (11 discharged, 10 under ICC team)

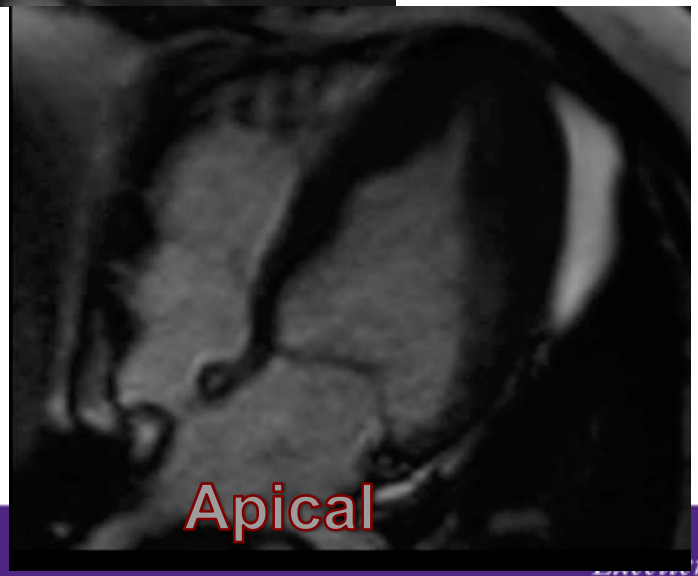
Mean 7.2 first degree relatives on Merseyside…..

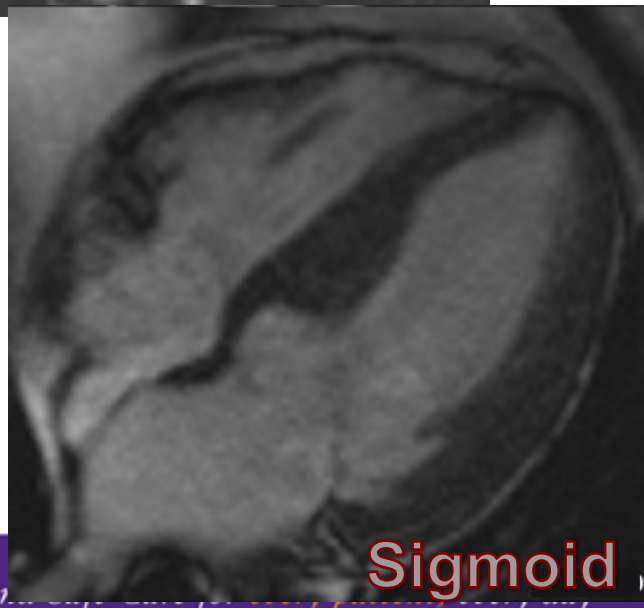
Significant breakdown in processes identified.

HCM Morphologies









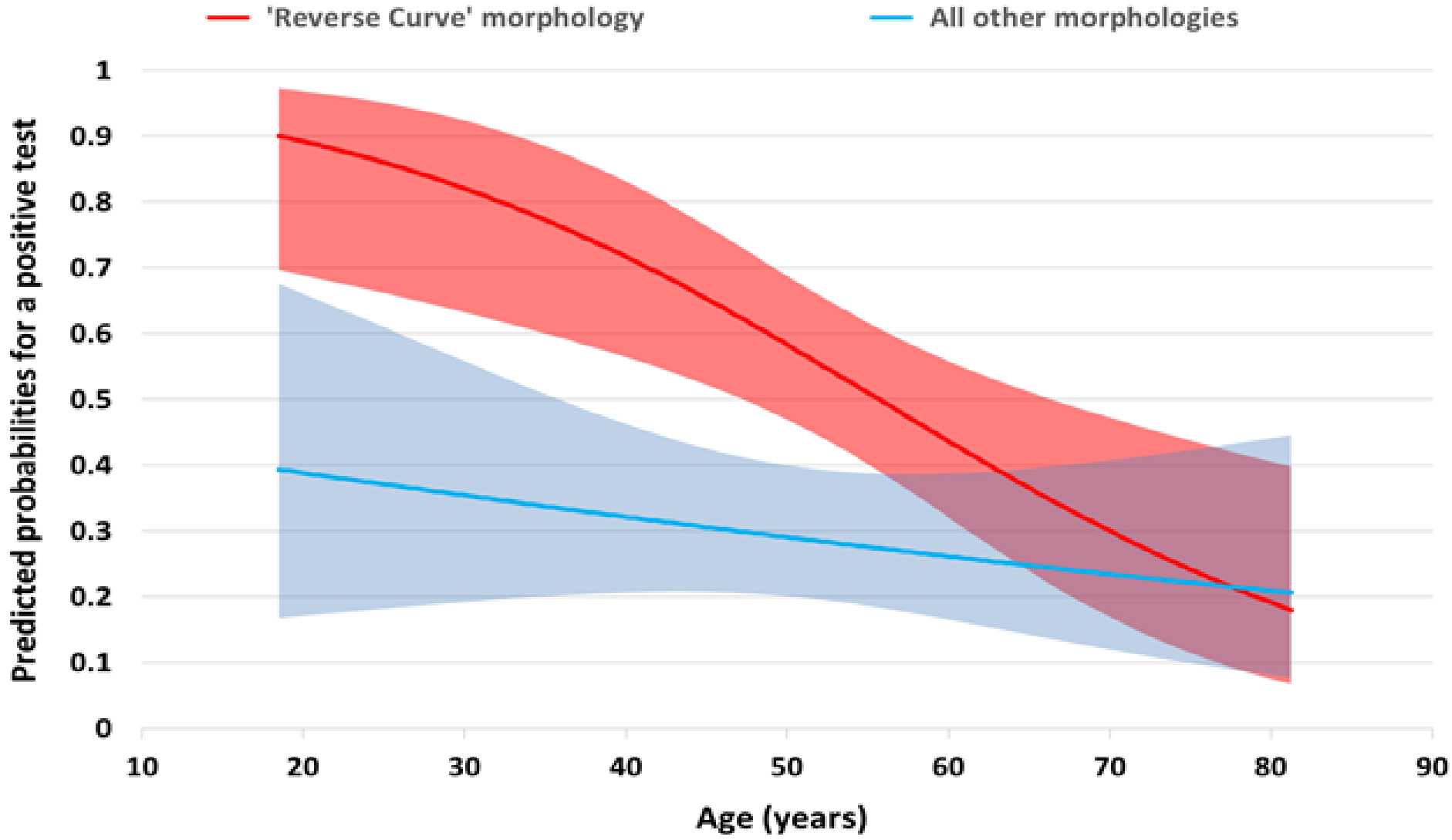
Clinical predictors of informative test

Aid decision making for genetic testing

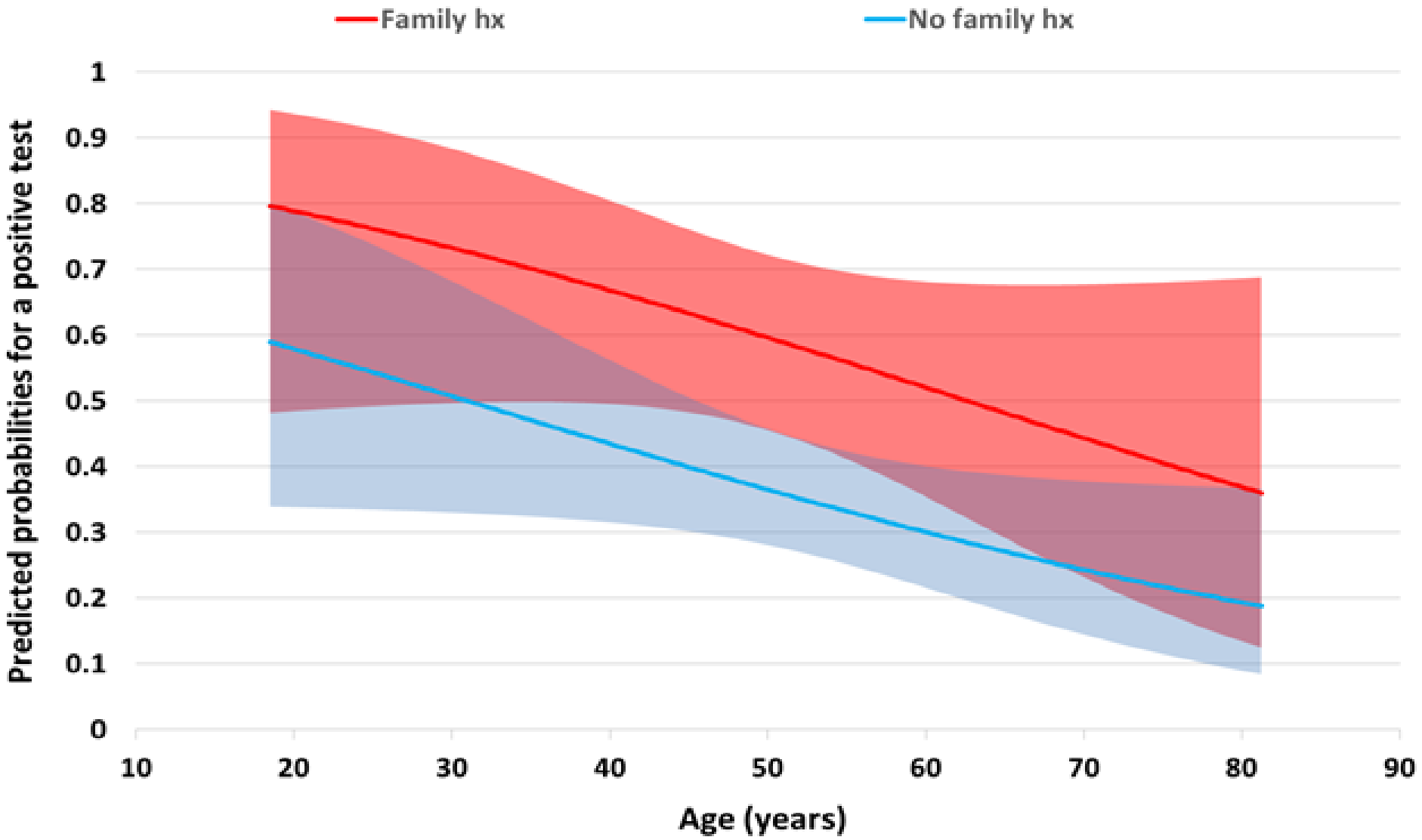
HCM panel costs £750-800

Younger age of diagnosis, family history of HCM or SCD, reverse curve morphology are positive predictors

Reverse curve and Younger Age



Family History and Younger Age



Clinical predictors of informative test

Multivariable logistic regression analysis		
	Odds Ratio (95% CI)	P value
Age at diagnosis (years)	0.97 (0.94, 0.99)	0.007
FH of SCD or HCM	2.18 (1.07, 4.42)	0.031
Reverse Curve	2.85 (1.45, 5.57)	0.002

Table 1: Multivariate regression analysis using predefined clinical characteristics

Likelihood of positive gene test using predictive model Factors: Age <50, FH SCD/HCM, reverse curve septal morphology	
No. Factors	Likelihood of positive result (%)
3/3	75.0
2/3	63.8
1/3	29.9
0/3	20.6

Table 2: The likelihood of an informative test using a predictive model

Which HCM patients should be tested

Index case with a clear clinical phenotype

WHO HAS

At-risk, first-degree family members that could be discharged from cardiac screening if predictive test negative

More severe phenotype = more likely +ve genetic test

Those with

- Family history HCM
- No known FH but reverse curve morphology and <50 years at presentation

Who not to test

Patient with no clear phenotype

Patient with no living relatives

In-patients, patients having procedures, patients in pacing clinic

Genetic testing is not indicated (in cardiology) to:

- Confirm all clinical diagnoses
- Diagnose a borderline or doubtful clinical diagnosis

Miles Frost Fund: Findings from the interim evaluation

Phase 2 : 8 sites over UK
Funding for 2 years given for Nurse Specialists, Genetic Counsellors, Co-ordinators, Admin support.

Provide the British Heart Foundation with a quarterly activity report

Results on diagnostic genetic testing in HCM show that the audit work previously undertaken reflects the results we see and we are now being selective of who we test.

LHCH – new approach to genetic testing

Liverpool Heart and Chest Hospital NHS
NHS Foundation Trust

Process	Role of the BHF nurse
Informative genetic test	
Proband referred to clinical genetics	Process all results to ICC email account MDT discussion of all results Template letter to GCs once satisfied can be used for predictive testing Contact proband to explain the results and rationale for review with GC Ensure pedigree is correct - contact details of relatives
Proband seen in clinical genetics Pedigree Dear family member' letters	Monthly check that referral has been processed Offer follow up appointment to proband for clarification Inform GP of the referral process for proband and relatives (template)
Relative seen in clinical genetics Genetic counselling Predictive testing	Receive referrals for those with a positive predictive test Arrange review in ICC clinics

ICC Nurse Specialist Role

- Education:
  - patient & family
  - Health Care Professionals – CMUK Clinical Advisory Group
    MedEd e-learning module HCM

- Communication – increasing awareness – collaboration with Charities
- AICC - Annual conferences
- Accessibility – phone line
- Clinical management – case load – diagnostic genetic testing - HCM
- Family screening – ensuring it happens (clinical or genetic)
- Facilitate Genetics & Imaging MDT
- Transition Clinic

ICC Team LHCH

- 2 dedicated ICC Cardiologists (1 spec echo 1 CMR, 1 septal reduction ASA)
- 2 device & Heart Failure Specialists– ICC
- 2 Imaging – Specialists
- 5 Electrophysiologists – device – ICC – 1 RF septal reduction
- 1 surgeon – surgical septal reduction - myectomy
- 1 Clinical psychologist
- 2 Genetic Counsellor sessions – (2 sessions admin – 2 full days funded Miles Frost
- 2 Geneticist sessions
- 2 ICC Nurse Specialist – 1 currently Miles Frost Funded
- 2 Co-ordinators
- 1 Typist

ICC Nurse Specialist Role Day to Day

Liverpool Heart and Chest Hospital NHS
NHS Foundation Trust

Monday	Tuesday	Wednesday	Thursday	Friday
Septal reduction MDT Nurse led Family Screening Clinic Provocation Testing	Admin - Letters Bi monthly Cardiomyopathy clinic	ICC Telephone clinic	Cardiomyopathy Clinic & Family screening Clinic	Prep & process MDT - Genetic MDT - Imaging 1/12 Transition clinic – Alder Hey Provocation Testing
Process Referrals	Septal reduction clinic Family screening clinic 1/12 Channelopathy clinic Bi monthly Cardiomyopathy clinic	Genetic Test results – process results	Admin –Results PM: Alternate months CMUK support group	Cardiomyopathy clinic Family screening clinic 1/12 HMD 1/12 Channelopathy clinic


THANK YOU FOR YOUR ATTENTION!
ANY QUESTIONS?