South Wales and South West
Congenital Heart
Disease Network

Paediatric High Dependency Network Day

Congenital Heart Disease Network for South Wales and South West

June 22nd 2017

Sheena Vernon

RGN MSc

Aims for the Day

- Background
- Standards
- Overview of the network

- Background
- Safe and sustainable, 2008
- Review of the provision of paediatric cardiac surgery services, consolidate practice
- Criteria for centres
- No ACHD
- Congenital heart Disease Standards and specifications NHS England 2015

The Standards

- Section A: The network approach
- Section B: Staffing and skills
- Section C: Facilities
- Section D: Interdependencies
- Section E: Training and education
- Section F: Organisation, governance and audit
- Section G: Research
- Section H: Communication with patients
- Section I: Transition
- Section J: Pregnancy and contraception
- Section K: Foetal diagnosis
- Section L: Palliative care and bereavement

NHS
England

Congenital Heart
Disease
Standards &
Specifications

Standard	Paediatric	Implementation timescale
Palliative Care Note: **Palliative care** is the active, total care of the patients whose disease is not responsive to curative or life-extending treatment.		
L1(L1)	Each Specialist Children's Surgical Centre must have a palliative care service able to provide good quality end-of-life care in hospital and with well-developed shared-care palliative services in the community which are appropriate to the physical, psychological, cognitive and cultural needs of the child/young person and family/carers. This must also include bereavement follow-up and referral for ongoing emotional support of the family/carers.	Immediate
L2(L1)	Clinicians should use nationally approved paediatric palliative medicine guidance to plan palliative care from the point of diagnosis.	Immediate
L3(L1)	When a child or young person is identified as needing palliative or end-of-life care, a lead doctor and named nurse will be identified by the multidisciplinary team in consultation with the child/young person and their family/carers. These leads may change over time as appropriate.	Immediate
L4(L1)	The lead doctor and named nurse will work together with the palliative care team to ensure the child/young person and their family/carers are supported up to, and beyond death.	Immediate
L5(L1)	An individualised end-of-life plan, including an advanced care plan, will be drawn up in consultation with the child/young person and their family/carers, and will include personal preferences (e.g. choice to remain in hospital or discharge home/hospice; presence of extended family). The potential for organ and tissue donation should be discussed. The family/carers and all the professionals involved will receive a written summary of this care plan and will be offered regular opportunities to discuss any changes with the lead doctor.	Immediate

Standard	Paediatric	Implementation timescale
L6(L1)	The lead doctor, with the named nurse, will ensure that the agreed end-of-life plan is clearly documented and agreed with all medical, nursing and psychological support team members (including lead clinicians in other treatment units and relevant community services) to ensure that all clinical staff understand the ongoing care and the reasons further active treatment may not be possible.	Immediate
L7(L1)	Communication and end-of-life care discussions with children, young people and their families/carers must be open, honest and accurate.	Immediate
L8(L1)	The child/young person and their family/carers must be offered details of additional non-NHS support services available to them.	Immediate
L9(L1)	For children and young people remaining in hospital, a named member of the nursing and medical staff will be identified during every shift so that they and their parents/carers can easily seek answers to questions and express wishes, worries and fears.	Immediate
L10(L1)	The room and environment must be prepared to meet the palliative care needs and wishes of the child/young person and their family/carers, and allow them the privacy needed to feel that they can express their feelings freely.	Immediate
L11(L1)	All members of the clinical team must be familiar with the bereavement services available in their hospital.	Immediate
L12(L1)	Children/young people and their families/carers must be made aware of multi-faith staff and facilities within the hospital.	Immediate
Discharge and out-of-hospital care		
L13(L1)	Any planned discharge must be managed by the named nurse who will coordinate the process and link with the child/young person and their family.	Immediate

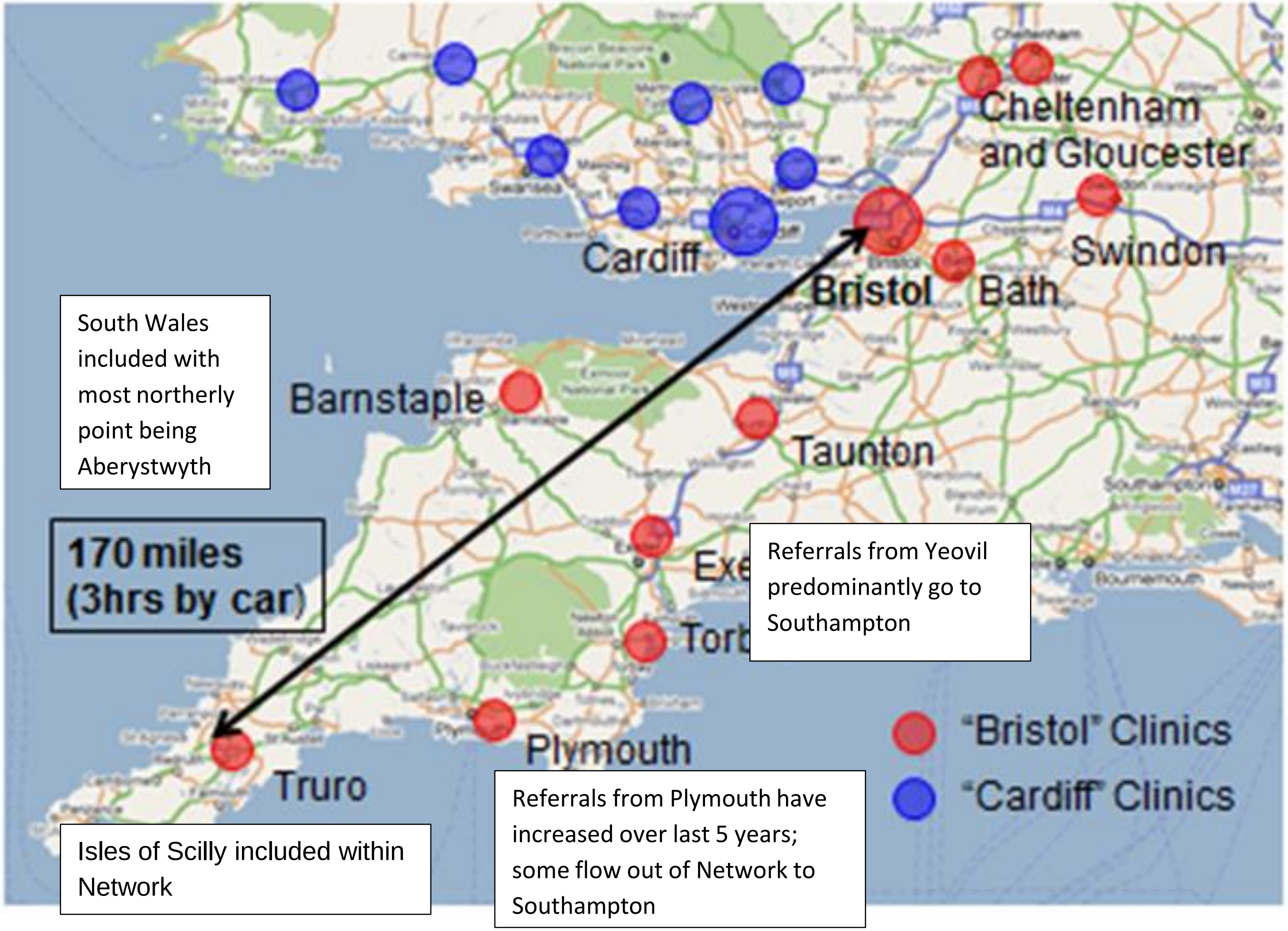
Standard	Paediatric	Implementation timescale
L14(L1)	The lead doctor, with the named nurse, will ensure that the end-of-life plan and discharge plan are shared with relevant community and local hospital services including local paediatricians, GPs, community children's nurses, out-of-hours GP and ambulance services and the local children's hospice. Written care plans must be provided for all members of the team. All equipment needed in the home must be available prior to discharge.	Immediate
L15(L1)	Support for children/young people and their families/carers must continue if they choose to have their end-of-life care in the community. Families/carers must be given written details of how to contact support staff 24/7. Community and outreach provision must be planned prior to discharge.	Immediate
	Management of a Death (whether expected or unexpected)	
L16(L1)	The team supporting a child/young person, and their family/carers, at the end of their life must adopt a holistic approach that takes into consideration emotional, cultural and spiritual needs, their ability to understand that this is the end of life, and must take account of and respect the wishes of the child/young person and their family/carers where possible.	Immediate
L17(L1)	If a family would like to involve the support of members of their home community, the hospital-based named nurse, as identified above, will ensure they are invited into the hospital.	Immediate
L18(L1)	Young people, parents and carers will be offered an opportunity to discuss the donation of organs and tissues with the *Donor* team.	Immediate
L19(L1)	The lead doctor/named nurse will inform the hospital bereavement team that a child is dying. They should only be introduced to the family/carers before a death has occurred, if they have specifically requested to meet them.	Immediate
L20(L1)	Families/carers must be allowed to spend as much time as possible with their child after their death, supported by nursing and medical staff, as appropriate. It is essential that families have an	Immediate

Standard	Paediatric	Implementation timescale
	opportunity to collect memories of their child.	
L21(L1)	When a death occurs in hospital, the processes that follow a death need to be explained verbally, at the family's pace and backed up with written information. This will include legal aspects, and the possible need for referral to the coroner and post-mortem. Where possible, continuity of care should be maintained, the clinical team working closely with the bereavement team. Help with the registration of the death, transport of the body and sign-posting of funeral services will be offered.	Immediate
L22(L1)	Informing hospital and community staff that there has been a death will fall to the identified lead doctor and/or named nurse in the hospital.	Immediate
L23(L1)	Contact details of agreed, named professionals within the paediatric cardiology team and bereavement team will be provided to the child/young person's family/carers at the time they leave hospital.	Immediate
L24(L1)	Staff involved at the time of a death will have an opportunity to talk through their experience either with senior staff, psychology or other support services, e.g. local bereavement support.	Immediate
Ongoing support after the death of a child/young person		
L25(L1)	Within one working week after a death, the specialist nurse, or other named support, will contact the family at a mutually agreed time and location.	Immediate
L26(L1)	Within six weeks of the death, the identified lead doctor will write to offer the opportunity for the family/carers to visit the hospital team to discuss their child's death. This should, where possible, be timed to follow the results of a post-mortem or coroner's investigation. The family/carers will be offered both verbal and written information that explains clearly and accurately the treatment plan, any complications and the cause of death. Families who wish to visit the hospital before their formal appointment should be made welcome by the ward team.	Immediate

Standard	Paediatric	Implementation timescale
L27(L1)	When a centre is informed of an unexpected death, in another hospital or in the community, the identified lead doctor will contact the family/carers.	Immediate
L28(L1)	If families/carers are seeking more formal ongoing support, the identified Children's Cardiac Nurse Specialist/named nurse will liaise with appropriate services to arrange this.	Immediate

- The Network

Team & Geography of the Network



South Wales included with most northerly point being Aberystwyth

170 miles (3hrs by car)

Referrals from Yeovil predominantly go to Southampton

Isles of Scilly included within Network

Referrals from Plymouth have increased over last 5 years; some flow out of Network to Southampton

Sheena Vernon, Lead Nurse

James Dunn, Network Manager

Dr Andrew Tometzki, Clinical Director

Network Vision

Our vision is to be a Network whereby:

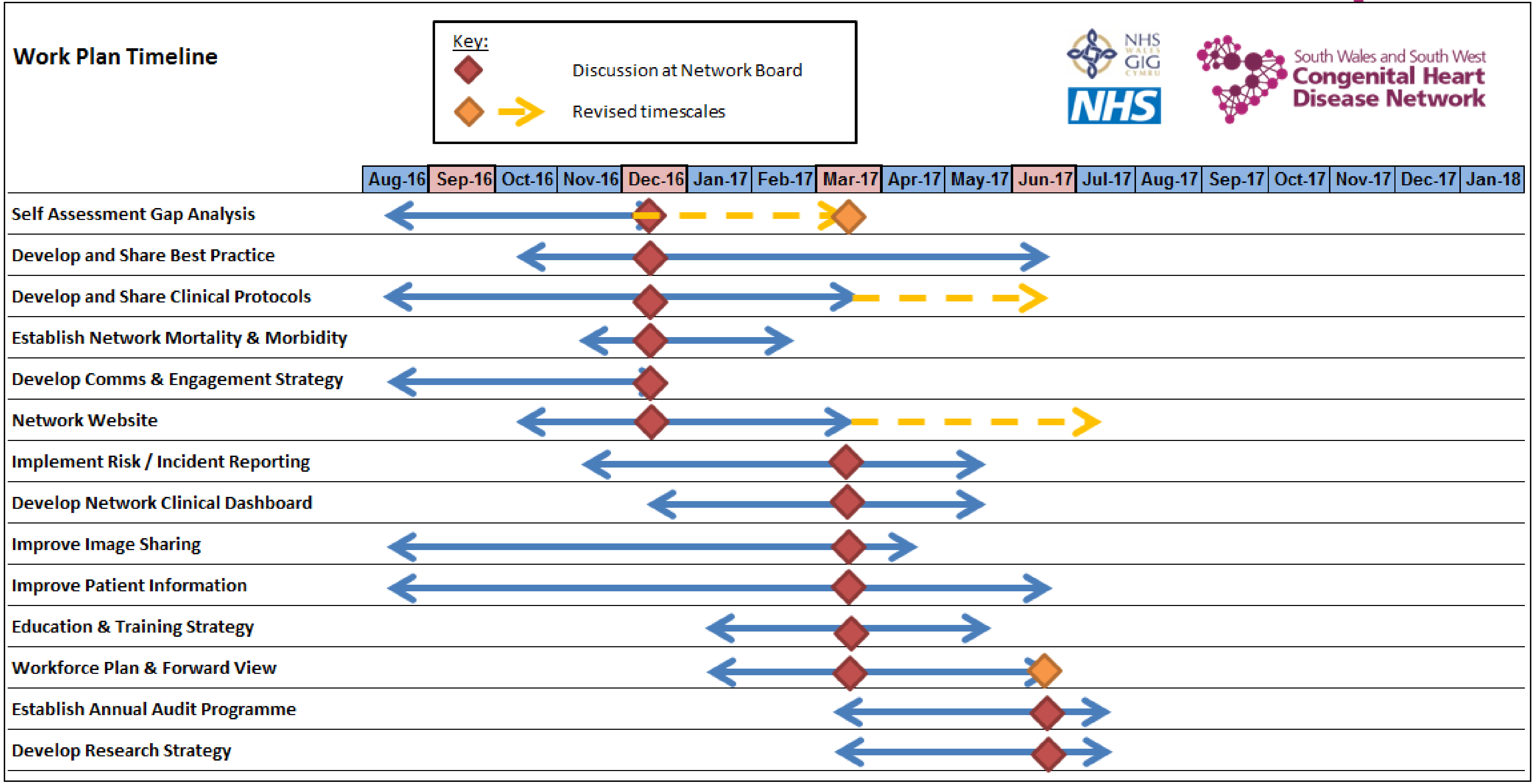
- Patients have **equitable access** to services regardless of geography
- **Care is provided seamlessly** across the Network and its various stages of transition (between locations, services and where there are co-morbidities)
- **High quality care** is delivered and participating centres meet national standards of CHD care
- The provision of **high quality information** for patients, families, staff and commissioners is supported
- There is a strong and **collective voice** for Network stakeholders
- There is a strong culture of collaboration and action to **continually improve** services

Funding and Accountability

- EOI submitted for 16/17 to commissioners to fund core Network team – **rejected**
- UHB therefore funding as well as hosting– but essential we operate as an **independent body** despite financial arrangement
- Longer term – this funding is **not sustainable**, **other models** will be explored
- Accountability will be to the **Network Board** (to be established) and ultimately NHS England's **Operational Delivery Network Oversight Board**

Network Work Plan Timings

South Wales and South West
Congenital Heart Disease Network



The Network Approach

•**sets out:** how networks will work
•**new/changing:** clear leadership (clinical and professional); cardiology (non-surgical) centres' participation in networks; second opinions and referrals

- **Challenge** : communication between local, cardiology and surgical centres
- CHD/CNS from SSC or SCS provide support, education and a link to network opd and ward staff
- Local link nurse in local centre/cardiac CNS +CHD

- Congenital heart disease

POPULATION

- Incidence: 8 per 1000 live births.
- 40 yrs. ago mortality from untreated CHD was 60%- 70% over the age of 18 years.
- Success of cardiac surgery and cardiology in infancy improved life expectancy.
- 85% of CHD patients, including complex, rare and severe conditions will reach adulthood.
- **More** adults than children with CHD.

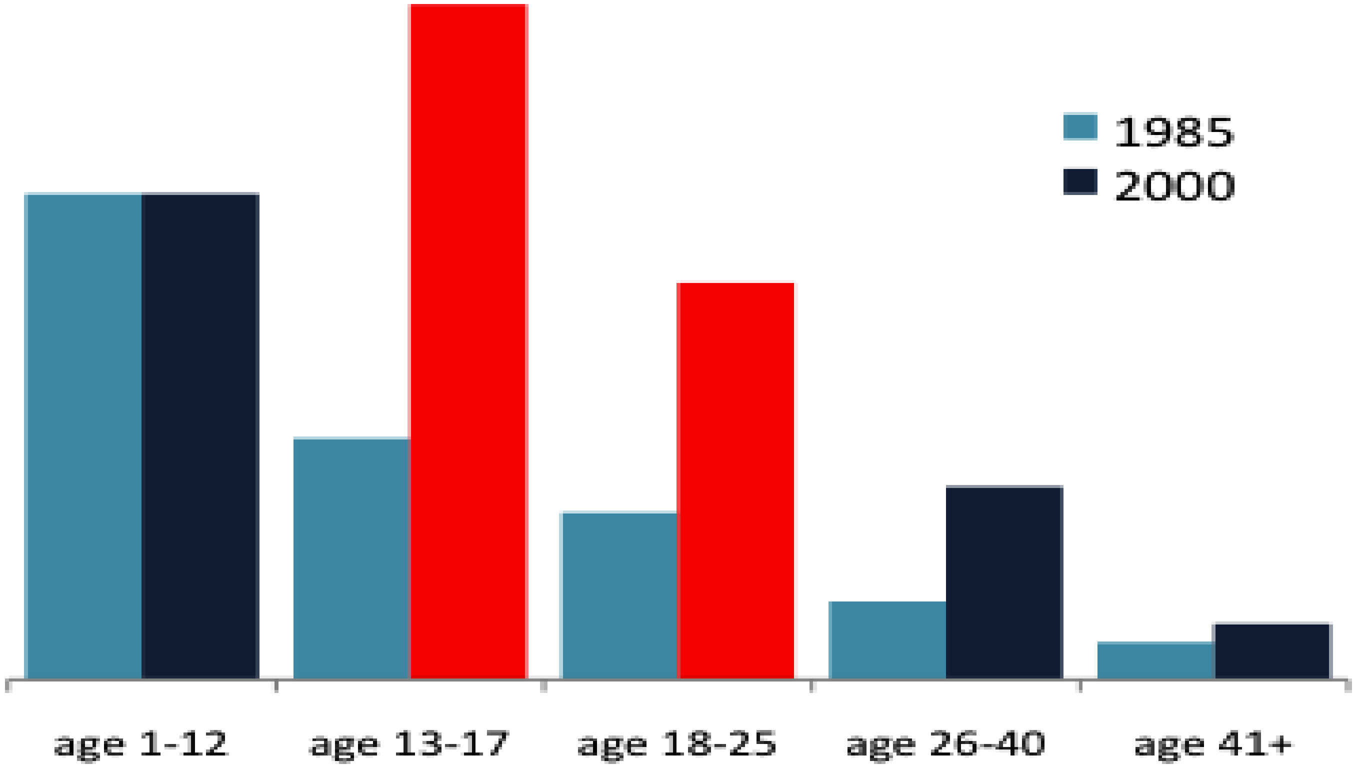
BCS 2002

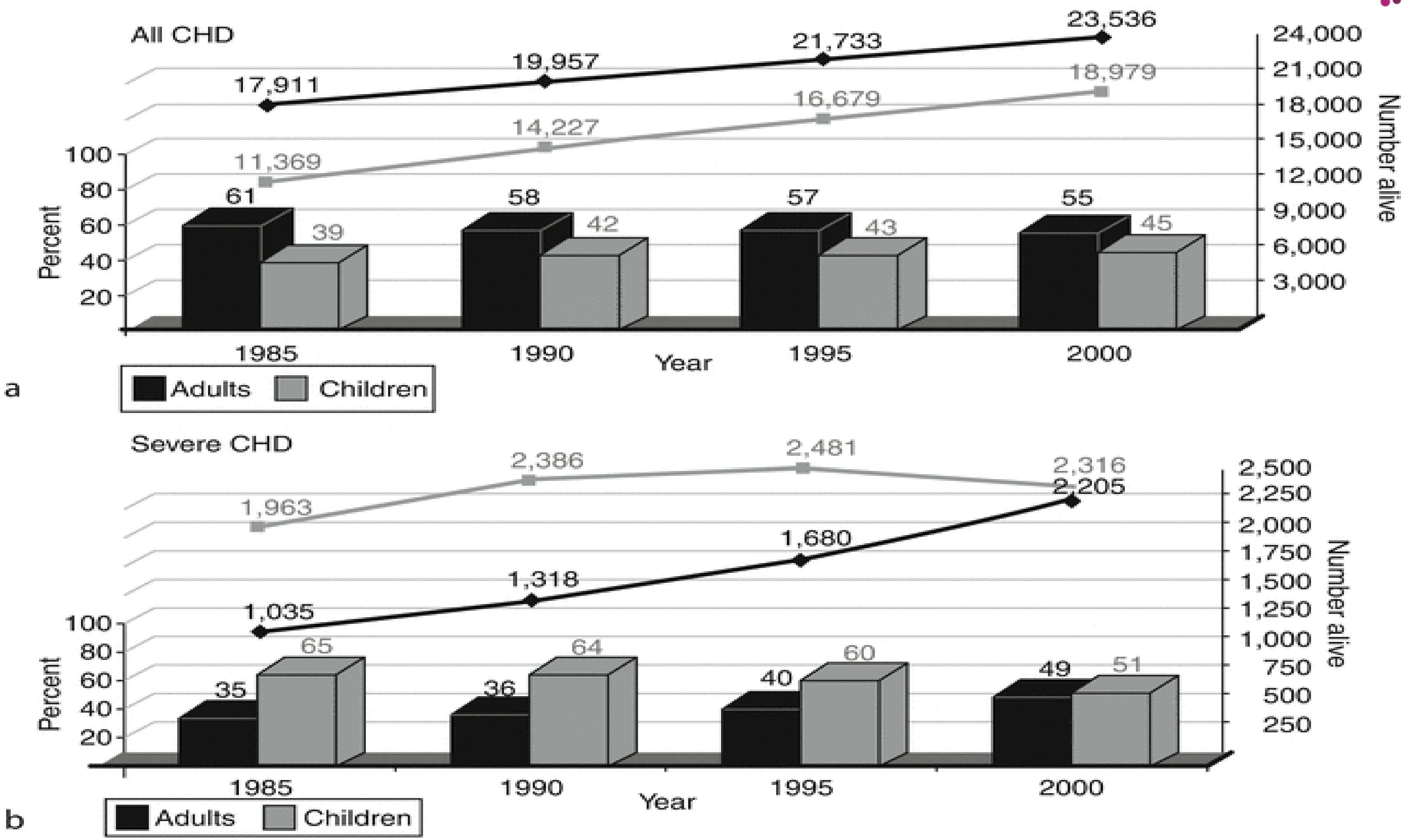
CURRENT POPULATION

- S.W England + parts of Wales 5 million.
- 7,000 **BHI.**
- 15,000 in South West.
- UK 160,000
- In 2000 equal numbers of those alive with severe CHD were adults.

Marelli A. J. et al 2007

number of 13-25 year olds increased x 3





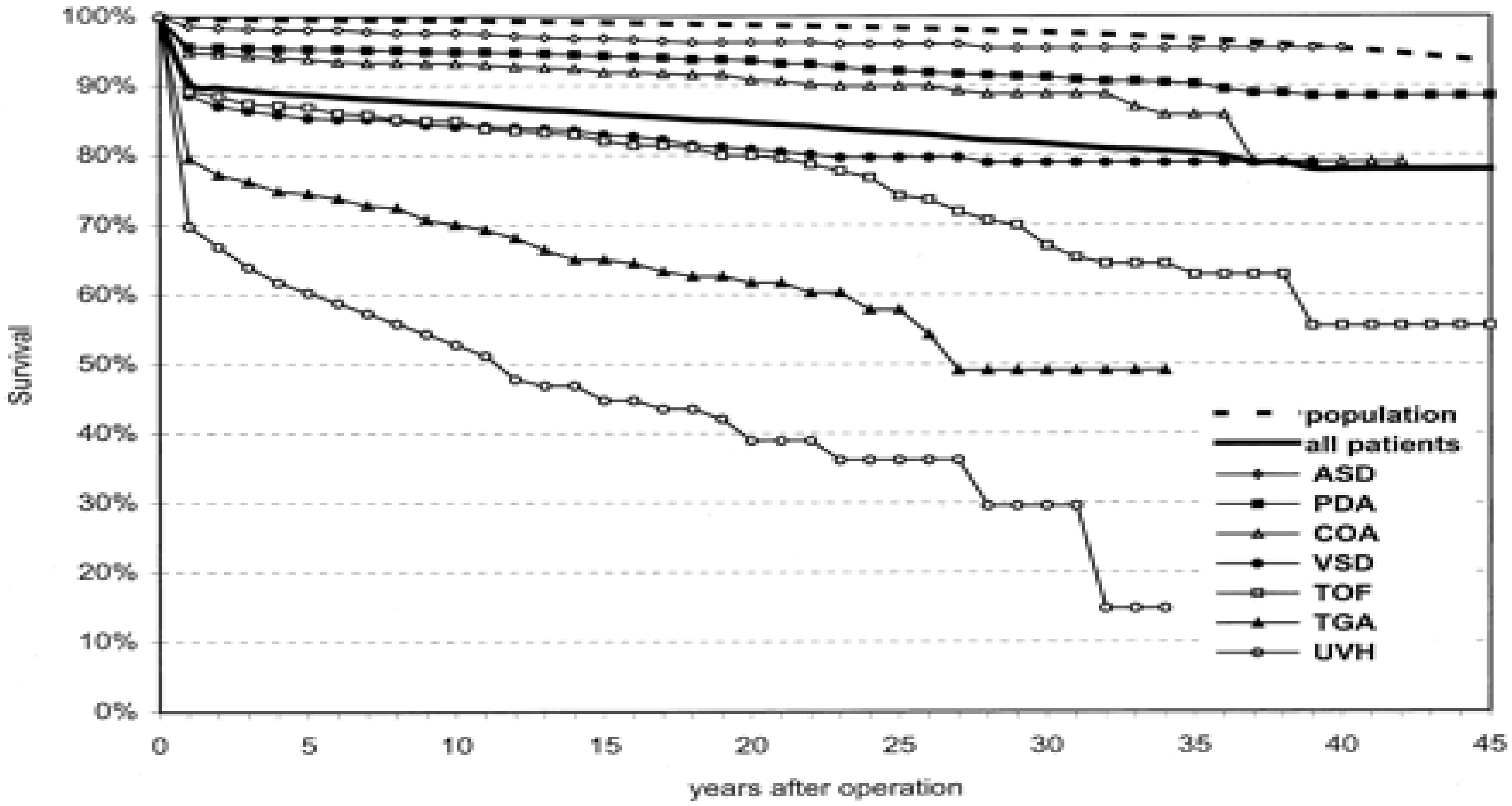
CHD = Congenital heart disease

From Marelli et all, J Amer Coll Card 2007

*Numbers and proportion of adults and children with all CHD (**a**) and severe CHD (**b**) in 1985, 1990, and 2000 (From Marelli et al. (2007) J Am Coll Card)*

LATE RESULTS OF PEDIATRIC CARDIAC SURGEY IN FINLAND

- Nieminen et al, Circulation 2001



Normal adolescence

- Time of physical and emotional development changes young people into adults, capable of self-care and independent decision
- Early (11-14yrs), middle (15-18years) and late (19-21)
- **Early adolescence** is identified by physical change and sexual maturation
- **Middle adolescence** there is a rapid growth of cognitive skills and understanding of abstract concepts. Peer groups play an increasing part in identity and separateness from parents
- **During late adolescence** a sense of self, sexual identity and an increased need for closeness, affection and intimacy forms
- Family life also undergoes a series of changes at this time
- *These normal individual and family developmental processes are greatly influenced by the context of chronic illness*

Tong and Kools 2004
Foster and Graham 2001

Role of CNS

- In-patient and out patient issues.
- Pre-assessment
- Surgery, cardiology, medical admissions, arrhythmias, endocarditis, heart failure.
- Learning disability work.
- Pregnancy/contraception.
- Teenage and young adult clinic.
- End of life care.
- Pulmonary hypertension.
- Telephone Advice >2000 calls pa.
- Patient information
- Education

COMMON CONGENITAL HEART DEFECTS

• Atrial Septal Defect	10%
• Ventricular Septal Defect	30%
• Tetralogy of Fallots	6%
• Transposition of the Great Arteries	4%
• Coarctation of the Aorta	7%
• Patent Ductus Arteriosus	10%
• Aortic Stenosis	6%
• Pulmonary Stenosis	7%
• Other	**20%**

Complex Lesions

- Cyanotic lesions
- Eisemnmengers Syndrome
- Ebsteins
- Hypoplastic left heart HLHS
- Hypoplastic right heart syndrome HRHS
- Tricuspid atresia
- TAPVD
- Cardiomyopathy
- Syndromes

- Causes of death

- Duct dependant
- Failure transplant

Palliative Care and bereavment

- UK children's heart surgery deaths have halved in 10yrs
- *Utley 2015*

Palliative Care and Bereavement

sets out: how to provide support at end of life and how to manage communication with families around the end of life
new/changing: all new

- **Challenge** : difficult conversations, patient, parents spouse, family and children
- Intense telephone advice
- End of life pathway
- Palliative care teams
- GP support

Add Paed Standards

Increasing Numbers

- **Increasing age of death into adulthood**
- Khairy P et al 2010 J Am Coll Cardiol

- **ACHD growing epidemic**
- Avila et al 2014 Canadian J Cardiol

- **Lifetime Prevalence of Congenital Heart Disease in the General Population From 2000 to 2010.**
- Ariane J. Marelli 2014;130:749-756; June 18, Circulation

Life Expectancy

- Reduced in severe CHD
- Improving - ICDs CRT
- **1/3** deaths sudden, others gradual
- EOL life, resus, pts wishes need to be discussed
- Avoids distress re: treatment decisions at EOL

Gatzoulis, Swan, Therrien, Pantely 2005

Care delayed/denied

- 48 patients, mean age 37yrs
- minority had EOL discussions prior to terminal admission
- most received aggressive treatment
- advanced palliative/EOL to
  - reduce hospital therapies
  - improve death and dying

Tobler D et al 2012 Pallliat Med

Call for early communication

- 200 pts + 48 CHD providers about EOL communication
- 2 patients + 50% providers discussed EOL
- 78% of patients wanted early discussion
- Drs discussions increase with complexity
- Early EOL discussion was favoured by 62% pts and 38% providers
- Earlier in disease course

Tobler D, 2012 Int J Cardiol

Knowledge and preference for ACP

- 200 pts
- 5% had completed, 56% never heard of them
- Infrequently completed in OPD
- HCP educate patients about advanced care directives
- Assist in preparing formal end of life planning
- Peoples wishes should be known and respected

Tobler et al 2012 www.ajconline.org

Now is the time to Act

- Balancing life prolonging intervention and EOL
- Parallel approach
- Learning from other specialities in managing young patients EOL
- Increasing numbers of severe ACHD , failing Fontans, decline inevitable
- Unique needs of young patients should be recognised.

Bowater S.E, SpeakmanJ.K. Thorne S.A (2013) Current Opinion in Supportive and Palliative Care.

Symptoms in EOL Cardiac disease

- Breathlessness, home O_2
- Anxiety, benzodiazepines, opiates, treat depression
- Anorexia, cachexia
- Profound lethargy
- Syncope (treatable rhythm disturbances)
- Refractory oedema (IV medication)
- Abdominal distention
- Early symptom control

Tobler D., De Stoutz N., Greutmann M(2011)
Current opinion in Supportive and Palliative Care

Avoid disaster

Discuss

Training

Resources


SOMETHING NEW
SAME OLD WAY

Help

Difficult Conversations

Making it easier to talk to people with heart failure about the end of life.

Funded by

British Heart Foundation

THE NATIONAL COUNCIL FOR PALLIATIVE CARE

Dying Matters

Written in with the National Council of Palliative Care

Aims to support those caring for people affected by heart failure

To open up conversations about end of life wishes and preferences

Help

Planning for your future care

A GUIDE

Preferred Priorities for Care

(Easy read)

the gold standards framework®

South Wales and South West
Congenital Heart
Disease Network

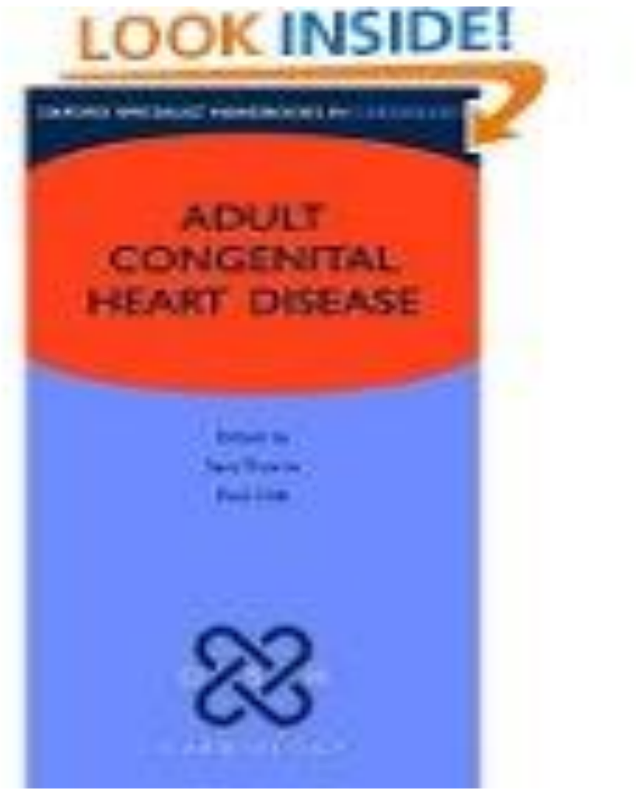
- The GSF Prognostic Indicator Guidance
- For clinicians to support earlier recognition of patients nearing the end of life
- The surprise question
- Next few months, weeks or days?

Cases

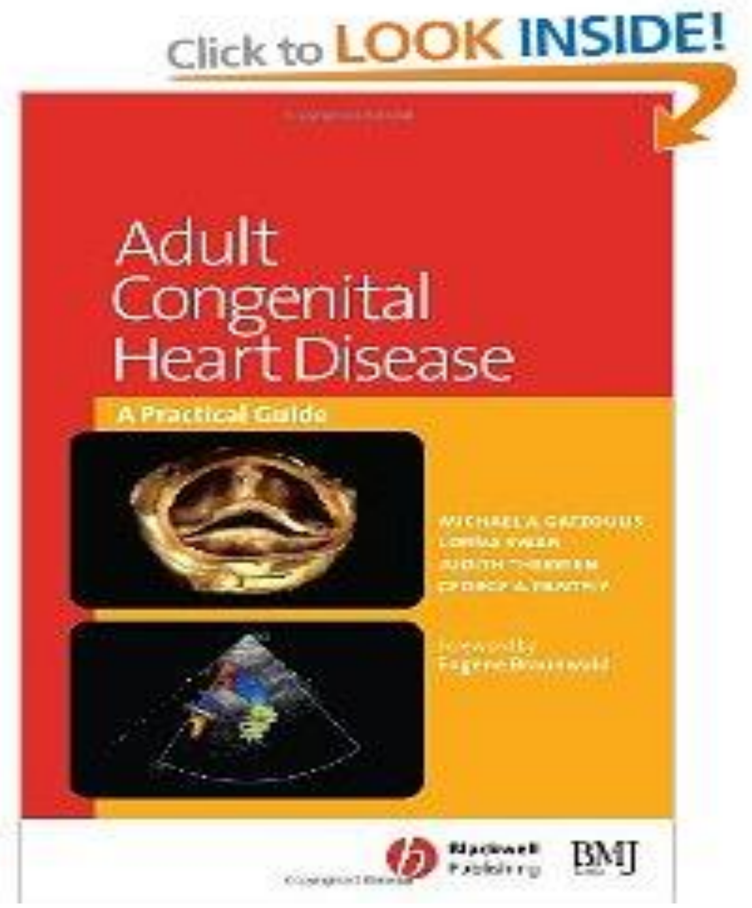
- K.A. 44 yrs post artificial heart
- M.B. 32yr old OOH
- R.B. 39yrs Welsh ICD
- A.L. 22yrs plus family
- A.D. 44yrs mild L/D
- S.H. 21yrs, PH palliative care

- Letter
- Family sometimes return
- No annual reminder
- Flowers to well known patients

Books


LOOK INSIDE!

ADULT CONGENITAL HEART DISEASE


Click to LOOK INSIDE!

Adult Congenital Heart Disease

A Practical Guide

BMJ