<table>
<thead>
<tr>
<th>Time</th>
<th>Session</th>
<th>Speaker</th>
</tr>
</thead>
<tbody>
<tr>
<td>08.30 - 09.00</td>
<td>Registration and refreshments</td>
<td></td>
</tr>
<tr>
<td>09.00 – 9.45</td>
<td>Overview of 8 common lesions</td>
<td>Sheena Vernon</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Lead Nurse</td>
</tr>
<tr>
<td>09.45 – 10.30</td>
<td>Surgery for the single ventricle</td>
<td>Mr Andrew Parry</td>
</tr>
<tr>
<td>10.30 – 11.00</td>
<td>Coffee</td>
<td></td>
</tr>
<tr>
<td>11.00 – 11.30</td>
<td>Red flags for general anaesthetic in complex congenital patients</td>
<td>Dr Amit Ranjan</td>
</tr>
<tr>
<td>11.30 - 12.00</td>
<td>Pregnancy with Fontan’s circulation</td>
<td>Dr Stephanie Curtis</td>
</tr>
<tr>
<td>12.00 - 12.30</td>
<td>Life threatening arrhythmia in Fontan patients</td>
<td>Dr Graham Stuart</td>
</tr>
<tr>
<td>12.30 - 13.30</td>
<td>Lunch</td>
<td></td>
</tr>
<tr>
<td>13.30 – 14.00</td>
<td>Fontan associated liver disease</td>
<td>Dr Radwa Bedair</td>
</tr>
<tr>
<td>14.00- 14.30</td>
<td>Patient story</td>
<td>Dr Andrew Shearn</td>
</tr>
<tr>
<td>14. 30- 15.00</td>
<td>Transferring complex patients to the adult services</td>
<td>Sheena Vernon RGN</td>
</tr>
<tr>
<td>15.00 – 15.15</td>
<td>Tea</td>
<td></td>
</tr>
<tr>
<td>15.15 – 16.00</td>
<td>Interactive cases with group quiz</td>
<td>Caryl Evans RGN</td>
</tr>
</tbody>
</table>
AIMS OF THIS TALK

• Develop knowledge and skills.
• Develop insight into on-going needs of the patient group & changes in their condition.
• Develop ability to provide information on services suitable for individual patients and family members.
• Enable patients to participate in decisions regarding health & social issues that arise throughout life.
OUTLINE

- Set the scene population
- Outline team
- What do we do
- Role of CNS
- Lesions
- Physical and psychological issues
- Guidelines
POPOPULATION

• 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.
With advances in pediatric cardiology and cardiac surgery, over 85% of children with congenital heart disease (CHD) now survive to adulthood. Studies estimate that there are approximately one million adults with CHD in the US and that this rapidly growing adult population probably outnumbers the children with CHD (Fig. 260.1). Early mortality and multiple morbidities, however, continue to affect these adults as they age.

**Figure 260.1** 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)

CHD = Congenital heart disease

From Marelli et al, J Amer Coll Card 2007
Numbers European Union (Population 497 Mill. in 2008)

- Prevalence 0.5 %* (2000)
- Prevalence 5.1 %* - 5.2 %** (2012/13)
- Extrapolation
- Estimated prevalence 11% (2030)

ACHD Patients > 60 years
ACHD Patients < 60 years
Children with CHD

** German Competence Network for Congenital Heart Disease (data on file)
RESULTS OF PEDIATRIC CARDIAC SURGERY IN FINLAND

Nieminien et al, Circulation 2001
Substantial improvements in outcome
## Relative age

<table>
<thead>
<tr>
<th></th>
<th>Patient's age (years)</th>
<th>Age difference:</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>20</td>
<td>25</td>
</tr>
<tr>
<td>ASD</td>
<td>25</td>
<td>26</td>
</tr>
<tr>
<td>Valvar disease</td>
<td>29</td>
<td>31</td>
</tr>
<tr>
<td>VSD</td>
<td>28</td>
<td>30</td>
</tr>
<tr>
<td>Aortic Coarctation</td>
<td>32</td>
<td>33</td>
</tr>
<tr>
<td>AVSD</td>
<td>33</td>
<td>34</td>
</tr>
<tr>
<td>Marfan syndrome</td>
<td>37</td>
<td>38</td>
</tr>
<tr>
<td>Tetralogy of Fallot</td>
<td>37</td>
<td>38</td>
</tr>
<tr>
<td>Ebstein anomaly</td>
<td>42</td>
<td>43</td>
</tr>
<tr>
<td>Systemic RV</td>
<td>46</td>
<td>48</td>
</tr>
<tr>
<td>Eisenmenger syndrome</td>
<td>57</td>
<td>58</td>
</tr>
<tr>
<td>Complex CHD</td>
<td>58</td>
<td>59</td>
</tr>
<tr>
<td>Fontan</td>
<td>64</td>
<td>65</td>
</tr>
</tbody>
</table>

Values present relative age adjusted for predicted 5-years mortality. Colors reflect the difference between relative and actual age. For example, a 40 year old Fontan patient has a mortality rate that is comparable to that of a 75 year old individual without CHD.
CURRENT POPULATION

- 8,000 Adults South West
- 6,500 Children 135,000 adults and young people England
- In 2000 equal numbers of those alive with severe CHD were adults.

Marelli A. J. et al 2007
CHD Network
CHD 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, audit
- Section G: Research
- Section H: Communication
- Section I: Transition
- Section J: Pregnancy contraception
- Section K: Fetal diagnosis
- Section L: Palliative care and bereavement
Welcome to the Congenital Heart Disease Network South Wales and South West

We proudly support over 6,500 children and 8,000 adults with a congenital heart condition.
Terminology

Grown-up Congenital Hearts (GUCH)

Adult Congenital Heart Disease (ACHD)
OUTPATIENTS
ADULT CONGENITAL TEAM

- BHI Cardiologists x 5, Surgeons x 3
- Specialist registrar, registrar x 2
- CNS x 3 (5)
- Obstetric team, 108 new pts
- Consultant Radiologists
- Anaesthetist CHD interest
- Peripheral clinics in 7 D.G.H’s
- Barnstable, Cheltenham, Swindon, Taunton, Exeter, Torbay, Truro
Role of ACHD CNS?

• In-patient and out patient issues
• Pre-assessment clinics
• 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
• Write patient information
• Education to pts and staff
<table>
<thead>
<tr>
<th>Condition</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>Atrial Septal Defect</td>
<td>10%</td>
</tr>
<tr>
<td>Ventricular Septal Defect</td>
<td>30%</td>
</tr>
<tr>
<td>Tetralogy of Fallots</td>
<td>6%</td>
</tr>
<tr>
<td>Transposition of the Great Arteries</td>
<td>4%</td>
</tr>
<tr>
<td>Coarctation of the Aorta</td>
<td>7%</td>
</tr>
<tr>
<td>Patent Ductus Arteriosus</td>
<td>10%</td>
</tr>
<tr>
<td>Aortic Stenosis</td>
<td>6%</td>
</tr>
<tr>
<td>Pulmonary Stenosis</td>
<td>7%</td>
</tr>
<tr>
<td>Other</td>
<td>20%</td>
</tr>
</tbody>
</table>
PREDISPOSING FACTORS

- Maternal drugs e.g. anti-epileptics, lithium, alcohol
- Chromosomal Aberrations e.g. 1 in 700 Downs Syndrome. 40% D.S. have C.H.D. Turners/Williams Syndrome
- Environmental Factors e.g. Radiation
- Infection/Virus e.g. Rubella
- Maternal Conditions
- e.g. Diabetes
Lesion information on all lesions on www.swswchd.co.uk

Professionals → Clinical information → adults
The Normal Heart

Superior vena cava

Aorta
Pulmonary artery

Pulmonary veins

Atrial septum

Right atrium

Pulmonary valve

Mitral valve

Aortic valve

Tricuspid valve

Left atrium

Left ventricle

Ventricular septum

Right ventricle

Inferior vena cava
Ventricular Septal Defect

- Increased blood flow to the lungs
- Enlarged right ventricle
- Enlarged left ventricle
- Ventricular septal defect
Coarctation of the Aorta

Coarctation

Thick left ventricle
Persistent Ductus Arteriosus

- Ductus
- Increased blood flow to the lungs
- Enlarged left ventricle
Aortic stenosis

Narrowed aortic valve
Pulmonary Stenosis

- Thickened Pulmonary valve
- Thick right ventricle
Tetralogy of Fallot

- Reduced pulmonary blood flow
- Large over-riding aorta
- Ventricular septal defect
- Narrow pulmonary valve
- Narrow outlet
- Thick right ventricle
Transposition of the Great Arteries

- Aorta from the right ventricle
- Pulmonary artery from the left ventricle
PHYSICAL ASPECTS OF CARE

- Infective endocarditis.
- Arrhythmias/heart failure.
- Surgery +/- re-operation – risks Intervention.
- Stroke.
- Cyanosis/Polycythaemia.
- Pregnancy/Contraception.
- Coronary Artery Disease.
INFECTION

ENDOCARDITIS

• Causes/risk?
• Diagnosis
• Bloods, TOE, ECG+ CXR
• Urine dip
• Treatment
• Complications
• Prophylaxis
• Nursing

www.nice.org.uk
Piercings
ARRHYTHMIAS

- Operative procedures from the early years, scarring affecting the conducting pathway.
- A/F, atrial flutter signs of deterioration in patients with Fontans, Fallots, A.S, single ventricle hearts and right sided conduit.
- Treatment return to S/R, anti-coagulate.
- Risk of S.C.D.
- Ablation, pacemaker or I.C.D.
- EOL discussion

www.heartrhythmmalliance.org/aa/uk
www.arrhythmiaalliance.org.uk
ARRHYTHMIA

- Urgent cardioversion
- Mapping
- Catheter ablation and surgical approaches
- Pacing/ICD
- Medication/side effects/pregnancy

- Danger Fontans and Ebsteins, TGA Mustards or Sennings flutter

- SVT most common
- VT in AS + TOF
RIGHT SIDED HEART FAILURE
(Cor Pulmonale)

- Fatigue
- ↑ Peripheral Venous Pressure
- Ascites
- Enlarged Liver & Spleen
- May be secondary to chronic pulmonary problems
- Distended Jugular Veins
- Anorexia & Complaints of GI Distress
- Weight Gain
- Dependent Edema
HEART FAILURE

- Medication ACE-inhibitors, angiotensin receptor blockers (ARBs)
- beta-blockers, aldosterone blockers (spironolactone or eplerenone)
- Diuretics, ivabradine, digoxin (occasionally)
- Fluid restriction, daily weight
- Lifestyle changes, smoking, diet, exercise, salt
- Devices, pacemakers, CRT, ICD
- Surgery, valve, LVAD, transplant
SURGICAL PROBLEMS

- Risk of re-operation in this group
- Adhesions, bleeding, longer by-pass time
- Renal and liver function problems
- Arrhythmias
- Cyanosed patient will require a higher PCV.
- Higher filling pressures needed in some conditions
- FBC
- Pericardial and pleural effusions
SURGICAL EMERGENCIES

• Complications
• Bleeding, infection, fever, thrombosis, embolism, fluid overload, dehydration
• Early detection vital
• Aggressive management
• Pain control for catecholamine stress
• Avoid early discharge
Cyanosis results from an increase in RBC as the body attempts to improve its oxygen carrying capacity.

- Increased viscosity, thrombosis, stroke, embolus, PH

- Caution if NBM, IV fluids
CYANOSIS
Watch for ……..

- Sepsis, brain abscess
- Renal function
- Gout
- Gall stones
- Orthopaedic complications
- Skin, acne, I.E.
- Ferratin
EMERGENCIES

- Arrhythmia
- Surgery
- Cyanosis
- Infection
- Ht Failure
- Ischaemia
- Pregnancy
- Transplant
PSYCHOLOGY

- Anxiety about heart condition, prognosis
- Repeated hospital visits
- Risk taking behaviour
- Compliance
- Depression
- Phobia
- L.D.

- Toolkits on website for patients [www.swswchd.co.uk](http://www.swswchd.co.uk)
Support

- Clinical experience in Level 1, mentoring across the network
- Education/ Study pack-link nurse resources
- Annual and regional study days
- National group BACCNA
BACCNA

• ‘British Adult Congenital Cardiac Nurses Association’

• Twice a year

• Agenda and networking

• BCCA November 19\textsuperscript{th}-20\textsuperscript{th} Newcastle
CHD 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, audit
- Section G: Research
- Section H: Communication
- Section I: Transition
- Section J: Pregnancy contraception
- Section K: Fetal diagnosis
- Section L: Palliative care and bereavement
To support adult standards guidelines from RCN for nursing published
READING
ESC Guidelines

ESC Guidelines for the management of grown-up congenital heart disease (new version 2010)

The Task Force on the Management of Grown-up Congenital Heart Disease (ESC)

Endorsed by the Association of European Paediatric Cardiology (AEPC)

Authors/Task Force Members: Helmut Baxingenauer (Chairperson) (Germany), Philippe Boudjemline (UK), Natalia M. S. de Groot (The Netherlands), Folke de Hennin (Germany), John A. Deanfield (UK), Nazarzadeh Saniei (Italy), Michael A. Gatzoulis (UK), Christian Goldblum-Berwald (Germany), Halsey Karmali (Germany), Philip Kuehn (UK), Roland Meijkmans (The Netherlands), Barbara J. M. Muleichik (The Netherlands), Brian O'Shields (Canada), Jose M. Oliver (Spain), Malek Scourby (France), Andrea Sestieri (Hungary), Erik Thuesen (Norway), Pascal P. Yudkin (France), Edmond Yomis (The Netherlands).

ESC Committee for Practice Guidelines (CPG): Alan Yagayek (Chairperson) (France), Angelo Artioli (Switzerland), Jerome Bas (The Netherlands), Claudo Bossi (Italy), Veronique Deau (France), Germaine Filippatos (Greece), Christian Frucht-Bronstein (France), Richard Hinds (UK), Peter Karmali (Australia), Thomas McDougal (UK), Bogdan A. Picewski (Romania), Zbigniew Reiter (Croatia), Udo Sichlmayr (Germany), Per Anton Simon (Sweden), Michel Tendler (Paraguay), Pim Varhoon (Greecce), Peter Waldorff (Czech Republic).

Document Coordinator: Giuseppe Ciccone (CPG Review Coordinator) (Italy), Uros Svet (Co-Reviewer Coordinator) (UK), Federico Andreucci (Italy), Maurice Bouchi (Switzerland), Maurizio Bregante (Italy), Andrea Bizzi (France), Stephan Broucker (UK), Werner Budts (Belgium), John Cross (Germany), Rafael de las Heras (Brazil), Gianna Johnson (France), Janna Kallio (Finland), Mara Kao (Greece), Alex Kassa (Turkey), Hadi Lee (Korea), Franco Moscato (Italy), Lavrete挪 (Portugal), Siddharth Naik (India), Robert Nosse (The Netherlands), Miguel Sousa Uva (Portugal), Pilar Terraza (Spain), Priya Triggle (Sweden), Patrizia Viennese (Italy), Harald Walschaerts (Korea), Hartmut Walter (UK), Gary D. Hiltz (USA), Jorgen Westerveld (Norway).

The disclosure forms of the authors and reviewers are available on the ESC website www.escardio.org/

ESC entities have participated in the development of this document:

Association for European Board of Cardiovascular Interventions (AEBI), European Heart Rhythm Association (EHRA), Heart Failure Association (HFA), European Association of Echocardiography (EAE), Congenital Cardiology, Council on Primary Care, Cardiovascular Imaging, Cardiovascular Nursing and Allied Professions (CCNP)

Working Groups: Grown-up Congenital Heart Disease, Pulmonary Circulation and Right Ventricular Function, Valvular Heart Disease, Cardiac Surgery, Thrombosis, Atrio-Ventricular Care.

ESC Guidelines for the management of grown-up congenital heart disease (new version 2010) was published as a supplement to the European Heart Journal (2010) 31, 2024–2168. For more information, please see the ESC guidelines website: www.escardio.org.
• European Society of Cardiology's guidelines on the “Management of Grown Up Congenital Heart Disease” 2010


• The 32nd Bethesda conference: Care of the Adult with Congenital Heart Disease JACC Vol 37, 2001.

• The Canadian Cardiovascular Society's Consensus Conference update 2001 update.
LOOKING AHEAD

- Congenital networks
- Support & encourage patients to lead as normal a life as their condition allows.
THANK YOU!

Questions?
MARFANS SYNDROME

- Tall and slender build
- Disproportionately long arms, legs and fingers
- Breastbone that protrudes outward or dips inward
- High, arched palate and crowded teeth
- Heart murmurs
- Extreme near-sightedness
- Abnormally curved spine
- Flat feet
MARFANS SYNDROME

• Connective tissue disorder, the heart (aortic dissection), eyes (dislocated lenses) and skeleton (scoliosis)
• Affects 1 in 5,000 births
• Reduced life expectancy in many patients
• **Cardiac manifestations** such as aortic dissection, aortic regurgitation and heart failure
• Cardiac surgery for abnormalities of the aorta
• Beta blockers
• [www.marfan.org.uk](http://www.marfan.org.uk)
GUIDELINES

Adult Congenital Heart Disease

A commissioning guide for services for young people and Grown Ups with Congenital Heart Disease (GUCH)
Team & Geography of the Network

Sheena Vernon, Lead Nurse

Caitlin Moss, Network Manager

Dr. Andrew Tometzki, Clinical Director

South Wales included with most northerly point being Aberystwyth

Isles of Scilly included within Network

Referrals from Yeovil predominantly go to Southampton

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

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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  
- **ACHD 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 +ACHD**
TRANSITION

- **sets out** seamless pathway of care to adult services
- **new/changing:** young people to be seen at least once at transition by a specialist with ACHD expertise; clear care plans/transition passports agreed; respecting particular needs of young people with *learning disabilities* and their carers.

- **Challenge:**
  - Big numbers
  - Young adult clinics, individual time + CNS time
  - Letters of introduction to patients
  - In-patient and out-patient support
  - Appropriate information
  - Avoid loss of F/up
Pregnancy

• Pre-pregnancy counselling for moderate to severe lesions & also:

• High risk, PH, severe Left sided lesions, Aortic root dilatation, cyanosis, ejection fraction less than 40%, mechanical valves.

• Care with ACE inhibitors, angiotensin11 receptors blockers and Amiodarone.
**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
49% adults in 2000
130 new pregnant referrals in 2013
Charities

- Newsletter / leaflets
- Telephone help line
- Support groups/mental health
- Financial support
- Workshops / conferences
- Web Sites
- BHF Lifestyle advice
Coaguchek machines

• INR test

• [www.roche-diagnostic.co.uk](http://www.roche-diagnostic.co.uk)

• [www.coaguchek.co.uk](http://www.coaguchek.co.uk)
Advice Line
Charities

- Newsletter / leaflets
- Telephone help line
- Support groups/mental health
- Financial support
- Workshops / conferences
- Web Sites
- BHF Lifestyle advice
PATIENT PHONE CALLS

- 2,000 calls pa admission, surgery, intervention, pregnancy, learning disability, TYA. Advice for HC professionals.
- Support, bereavement.
- Long haul flights/ travel.
- Employment issues/benefits.
- Managing Warfarin – INR – Coagu check.
- Tel. Pre-op.
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• 2,000 calls pa admission, surgery, intervention, pregnancy, learning disability, TYA. Advice for HC professionals.
• Support, bereavement.
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Caitlin Moss, Network Manager
Dr Andrew Tometzki, Clinical Director
ACHD

Structural
Intervention

PFO and
stroke disease

Pulmonary
Hypertension

Obstetric
Cardiology

Inherited
Cardiac
Conditions
Learning Disabilities

- Increasing numbers of patients having procedures and treatment
- 1 in 700 born with Downs, 40% will have CHD
- Time consuming
- Support for patient, family, CLDT and carers
- Capacity to consent? Best interest meetings?
- Appropriate communication
NURSING TEAM OF THE YEAR
2014
Lifestyle issue

- Outline of population
- Diet, alcohol, smoking and drugs
- Endocarditis
- Exercise
- Sex, pregnancy and contraception
- Extreme sport
- Risk taking
- Travel
- Support
Arrhythmias

- Causes
- Precipitating factors
- Deterioration
- Treatment
- Structural v Electrical
- Haemodynamics
- SVT most common
- VT in AS + TOF
ADVICE LINE
number of 13-25 year olds increased x 3
2007-2014

Letters Received

POAC

Calls