ADULT CONGENITAL HEART DISEASE
AN OVERVIEW

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Adult Congenital Heart Unit
Bristol Heart Institute
2018
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.
• Set the scene population
• Outline team
• What do we do
• Role of CNS
• Lesions
• Physical and psychological issues
• Guidelines
• 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.
• Average life expectancy 49 years
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.

**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)**
RESULTS OF PEDIATRIC CARDIAC SURGERY IN FINLAND

Niemininen et al, Circulation 2001
# Relative age

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.
Numbers European Union (Population 497 Mill. in 2008)

Prevalence
0.5%* (2000)

5.1%* - 5.2% ** (2012/13)

ACHD Patients < 60 years

ACHD Patients > 60 years

Children with CHD

Estimated prevalence 11% (2030)

** German Competence Network for Congenital Heart Disease (data on file)
number of 13-25 year olds increased x 3
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*
WHO KNOWS?!

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
- Obstetric team, 108 new pts
- Consultant Radiologists
- Anaesthetist
- Peripheral clinics in 7 D.G.H’s
- Barnstable, Cheltenham, Swindon, Taunton, Exeter, Torbay, Truro
WHAT DO NURSES DO?

- 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
ADVICE LINE
<table>
<thead>
<tr>
<th>Congenital Heart Defects</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
MARFAN'S 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)
MARFANS SYNDROME

• Tall and slender build.
• Disproportionately long arms, legs and fingers.
• A breastbone that protrudes outward or dips inward.
• A high, arched palate and crowded teeth.
• Heart murmurs.
• Extreme near-sightedness.
• An abnormally curved spine.
• Flat feet.
**Ventricular Septal Defect**

- **Increased blood flow to the lungs**
- **Enlarged right ventricle**
- **Enlarged left ventricle**
- **Ventricular septal defect**

Diagram illustrates a heart with a septal defect allowing blood to shunt from the right to the left side of the heart, increasing blood flow to the lungs and causing an enlarged right 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.
Piercings
INFECTION

ENDOCARDITIS

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

www.nice.org.uk
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.heartrhythmalliance.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 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
• Risk of re-operation in this group.
• Adhesions, bleeding, longer by-pass time.
• Affects renal and liver function.
• Risk of arrhythmias
• Cyanosed patient will require a higher PCV.
• Higher filling pressures needed in some conditions FBC
• Pericardial and pleural effusions may occur.
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
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.
Support

- Clinical experience in BHI
- Mentoring across the network
- Education/ Study pack
- Annual study days
- Regional training days
- National group BACCNA
- Support
BACCNA

• ‘British Adult Congenital Cardiac Nurses Association’

• Twice a year

• Agenda and networking

• BCCA November 14th
LOOKING AHEAD

• Congenital networks

• Support & encourage patients to lead as normal a life as their condition allows.
To support adult standards guidelines from RCN for nursing published
READING
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: European Association for Cardio-Thoracic Surgery (EACTS), the European Association of Nuclear Medicine (EANM)

Authors/Task Force Members: Gilbert Habib* (Chairperson) (France), Patrizio Lancellotti† (co-Chairperson) (Belgium), Manuel J. Antunes (Portugal), Maria Grazia Bonocore (Italy), Jean-Paul Cadiere (France), Francesco D’Avolati (Italy), Raúl Daiguérou (Belgium), Gebriné El Khoury (Belgium), Paola Anna Erba* (Italy), Bernard Jung (France), Jose M. Miro* (Spain), Barbara J. Muirder (The Netherlands), Edyta Pierska-Gosciniak (Poland), Susanna Price (UK), Jochen Roes-Hesselfink (The Netherlands), Ulrika Sverg-Martins (Sweden), Frank Tshuny (France), Pilar Torres Mas (Spain), Istrate Vilacosta (Spain), and Jose Luis Zamorano (Spain)

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Adult Congenital Heart Disease

A commissioning guide for services for young people and Grown Ups with Congenital Heart Disease (GUCH)
GUIDELINES

- 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.
THANK YOU!

Questions?
Team & Geography of the Network

Referrals from Yeovil predominantly go to Southampton.

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

South Wales included with most northerly point being Aberystwyth.

Isles of Scilly included within the Network.

Sheena Vernon, Lead Nurse

Caitlin Moss, Network Manager

Dr Andrew Tometzki, Clinical Director
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
**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**
• **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
- www.coaguchek.co.uk
Advice Line
Charities

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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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Piercings
MARFANS SYNDROME
Team & Geography of the Network

South Wales included with most northerly point being Aberystwyth

Isles of Scilly included within Network

170 miles (3hrs by car)

Referrals from Yeovil predominantly go to Southampton

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

Sheena Vernon, Lead Nurse

Caitlin Moss Network Manager

Dr Andrew Tometzki, Clinical Director
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