Stroke in Adults with Congenital Heart Disease: Incidence and Cumulative Risks

Sara Thorne
Impact of Stroke on Young Adults

- Death
- Major morbidity
- Loss of independence
- Loss of exercise capacity
- Psychological
- Socioeconomic
  - academic, career, earning potential
  - relationships
  - ability to support dependents
Adults Survivors of Congenital Heart Disease and Stroke Risk

**ACHD Stroke Risk**

**Childhood Risk**
- Perioperative stroke
- Right to left shunts – paradoxical embolism
- Prothrombotic circulations
- Mechanical valves
- Anticoagulation
- Arterial dissection
- Sickle cell disease
- Co-existent neurovascular lesions
- Focal arteriopathy of childhood

**General Adult Population Risk**
- Arrhythmia
- Hypertension
- Hyperlipidaemia
- Chronic kidney disease
- Heart failure
- Diabetes
- Peripheral vascular disease
- Smoking
- Obesity
- Alcohol
Causes of Stroke in Adults with Congenital Heart Disease

**EMBOLIC**
- Arrhythmia
- Endocarditis, cerebral abscess
- Devices
- Prosthetic valves
- Percutaneous procedures
- Watershed - hypoperfusion

**ISCHAEMIC**
- Hypertension

**PARADOXICAL EMBOLISM**
- Pulmonary AVM
- Veno venous collaterals
- Perforate atrial septum
- Palliated/unoperated cyanotic disease

**HAEMORRHAGIC**
- Aneurysm/ AVM
- Surgical anticoagulation
- Chronic anticoagulation

University Hospital
Birmingham
NHS Foundation Trust
Substrates for Arrhythmia in Adults with Congenital Heart Disease

- Surgical scars
- Atrial dilatation - volume loading current – new, chronic sequelae from early life septal defects regurgitant valves
- Fibrosis & hypertrophy 2° to chronic haemodymanic stress
- Heart failure
- Fontan circulation
- ....all the ‘acquired’ risk factors
How Common is Stroke in Young People?

Etiology and Treatment of Arterial Ischemic Stroke in Children and Young Adults

Review – general population
- Incidence of ischaemic stroke per 100 000

<table>
<thead>
<tr>
<th>Age Group</th>
<th>Incidence per 100 000</th>
</tr>
</thead>
<tbody>
<tr>
<td>Children (&lt;18)</td>
<td>1.2-2.4</td>
</tr>
<tr>
<td>Young adult (18-50)</td>
<td>3-11</td>
</tr>
<tr>
<td>Older adult (&gt;50)</td>
<td>88-149</td>
</tr>
</tbody>
</table>

Curr treatment options neurol 2014 Poisson et al

Childhood Stroke and Congenital Heart Disease

Laurence Ducharme-Crevier, MD1,2 and Mark S. Wainwright MD, PhD1

- Case control study
  - 2.5 million children
  - 412 stroke – 4% had CHD

- Children with CHD & surgery - 31 x ↑ risk of stroke compared to normal controls
Swedish register
- 26,000 born 1970-1993
- 10 matched controls each
- Ischaemic stroke 140 (0.5%)

- Stroke risk 11x > general population
- Risk factors
  - heart failure
  - hypertension
  - AF

Cumulative incidence of ischaemic stroke
30,000 ACHD patients age 18-64, 1998-2010

- Cumulative risk of ischaemic stroke for an 18 year old up to age 64
  - men 7.7% - 1 in 11
  - women 6.1% - 1 in 15

- Compared to normal population
  - risk of ischaemic stroke 9-12 times ↑ age <55
  - 2-4 times ↑ age 55-64
Cumulative stroke risk by lesion type

Predictors of ischaemic stroke

- Heart failure
- Diabetes
- Recent MI
- Lesion type
Importance of Atrial Arrhythmia as a Risk Factor for Stroke?

- Is subclinical arrhythmia an important stroke risk factor?

- Population / registry studies -
  - Dependent on accurate coding
  - Are subclinical arrhythmias included?
    - what kind of surveillance?
      - ...5 yearly 24 hour tape?
      - ...Yearly 24, 48, 72 hour tape?
      - ...Implantable loop recorder?

- Anticoagulation
  - Who?
  - How?
Incidence of Atrial Fibrillation in the General Population

The Clinical Profile and Pathophysiology of Atrial Fibrillation
Relationships Among Clinical Features, Epidemiology, and Mechanisms

Jason Andrade, Paul Khairy, Dobromir Dobrev, Stanley Nattel (Circ Res. 2014;114:1453-1468.)

- Review of general population studies of AF

**Incidence** of AF per 1000 person years
- <50y = <2
- By 60y = 5
- By 70y = 12
- By 80y = 25

Figure 2. A comparison is made of the atrial fibrillation (AF) incidence rates from published studies. In cases in which the studies report on sex-specific incidence by age group, the age group average is used for purposes of comparison. The thick line represents average AF incidence rates by age group, as derived from a pooled analysis of the individual studies weighted by sample size.
Incidence of Atrial Fibrillation in the General Population

The Clinical Profile and Pathophysiology of Atrial Fibrillation
Relationships Among Clinical Features, Epidemiology, and Mechanisms

Jason Andrade, Paul Khairy, Dobromir Dobrev, Stanley Nattel (Circ Res. 2014;114:1453-1468.)

- Prevalence – represents burden
- ACHD patients have this risk
  AND the risks peculiar to their disease

---

**Prevalence** of AF

- <50y = <1%
- By 60y = 2%
- By 70y = 5%
- By 80y = 12%

---

Figure 1. A depiction of the atrial fibrillation (AF) prevalence distribution found by each study published to date. This depiction uses the sex-specific average rates of AF prevalence, grouped by age. The thick line represents average AF prevalence rates by age group, as derived from a pooled analysis of the individual studies weighted by sample size.
Burden Of Atrial Arrhythmia And Stroke In ACHD – Birmingham Experience

- 4000 ACHD records, 2000-2015
- 347 (8%) atrial arrhythmia, mean age 49y
- 12% thromboembolic event, majority stroke
- Mean CHAD\textsubscript{2}DS\textsubscript{2}VASC score 1.08
- 19% anticoagulated at time of thromboembolic event

- High prevalence of atrial arrhythmia at young age in ACHD population
- CHAD\textsubscript{2}DS\textsubscript{2}VASC appears to be poor predictor of stroke
• Retrospective multicentre study, 5 year follow up
N= 199 patients with AF
Mean age 1\textsuperscript{st} AF 49 years

• Stroke occurred in 13%
50\% stroke before diagnosis of AF

Onset AF younger than general population
Stroke common & often predates diagnosis of AF

Need \textuparrow vigilance to detect AF
Prevalence of atrial tachyarrhythmia in adults after Fontan operation

Emily Quinton,1 Peter Nightingale,2 Lucy Hudsmith,3 Sara Thorne,3 Howard Marshall,1 Paul Clift,3 Joseph de Bono1

Single centre, n=166

Risk factors for arrhythmia

- ↑ age at Fontan
- Time since Fontan .... inevitable by 25 years?
- NOT – type of Fontan
Conclusion – Stroke In Adults With Congenital Heart Disease

- Stroke has a major impact on this ageing population
  Sequelae of stroke in early life
  Continuing risk of new stroke throughout adulthood due to ‘normal’ ageing
  lesion specific risk

- Atrial arrhythmia is a major - modifiable - risk factor
  How hard should we look for occult arrhythmia?
  Who should we anticoagulate?