Sub-aortic stenosis in England 1997 – 2015:
Reoperation rates and risk factors


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The *textbook* introduction

- “A description given to a spectrum of lesions resulting in fixed LVOTO”.

- *Not* dynamic forms of LVOTO e.g. HOCM.

- Relatively common:
  - 6.5% of adult congenital heart disease; 14% of LVOTO.

- Commonly associated with other forms of CHD (≈60%).

- Spectrum of disease:
  - Discrete subvalvular ring vs. long fibromuscular tunnel;
  - Progresses at a variable rate.
Editorial Note

Fixed subaortic stenosis – a frequently misunderstood lesion *

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The reality

• It may not be *that* simple.
• Is it even congenital?

• The pathological initiator is likely to reside in the myocardium.

• The mechanism by which the abnormal hypertrophic response within the LVOT is generated is as yet unclear.

Cape E.G. *et al*. JACC, 30(1), 247-254.
Introduction/background

• The timing and type of intervention remains controversial.

• Long-term survival is felt to be excellent, but there is a substantial incidence of recurrence of stenosis and late re-operation, as well as the development of aortic regurgitation.

• Long-term outcome data, when available, are limited to single centre or single surgeon experiences with small patient numbers.

• Population-level data to inform the management, risk stratification and follow-up of SubAS patients are lacking.
Methods

• Retrospective analysis of the ‘Hospital Episode Statistics’ data set for England from 1997 to 2015.

• SubAS patients admitted to hospital were identified using ICD-10 code “Q24.4”.

• Patients who underwent procedures for the treatment of SubAS were selected (OPCS-4 “K24.5-7”, “K31.2”, “K32.2”, “K35.2”, and “K37.3”).

• Where possible, CHD was classified as “simple”, “moderate” or “complex”; the latter group were excluded.

Statistical analysis

• Kaplan-Meier method was used to assess intervention-free survival following the first recorded procedure in the entire population and in subgroups.

• Survival between groups was compared using the log-rank test.

• Association of variables with survival was assessed using a multivariable Cox proportional hazards model.

• A p-value of < 0.05 indicated statistical significance.
Results

- 3,113 patients with a primary diagnosis of SubAS were identified.
- 68 patients were excluded due to incomplete or inconsistent data. Data from 3,045 patients were analysed.
- Median age at first repair was 7.6 [0-84.6] years.
- The majority were male (57.8%).
- 1770 (58.1%) of 3045 patients had an associated congenital heart defect.
Current study | Meta-analysis+  
--- | ---  
| | All patients | Natural history | Surgery |  
Parameter | Unit | n = 3,045 | n = 809 | n = 1476 |  
Male gender | % | 1799 | 57.8% | 60.6% | 59.2% |  
Age at first intervention | Years | 7.6 [0-84.6] | 7.7 (7.6-7.9) | 8.0 (7.8-8.1) |  
0-1y | n / % | 94 | 7.0% | - | - |  
1-18y | n / % | 974 | 72.6% | - | - |  
>18y | n / % | 274 | 20.4% | - | - |  
Associated CHD |  
Bicuspid aortic valve | n / % | 353 | 11.6% | 10.7% | 25.1% |  
Co-arctation of the aorta | n / % | 353 | 11.6% | 14.0% | 12.8% |  
Ventricular septal defect | n / % | 729 | 23.9% | 21.9% | 20.6% |  
Atrial septal defect | n / % | 257 | 8.4% | 5.9% | 6.6% |  
Aortic regurgitation | n / % | 157 | 5.2% | - | - |  
Mixed aortic valve disease | n / % | 24 | 0.8% | - | - |  
Valvular aortic stenosis | n / % | 315 | 10.3% | - | - |  
Supra-valvular aortic stenosis | n / % | 57 | 1.9% | - | - |  
Mitral valve disease | n / % | 326 | 10.7% | - | - |  
Atrioventricular septal defect | n / % | 148 | 4.9% | - | - |  
Patent ductus arteriosus | n / % | 270 | 8.9% | - | - |  
Pulmonary stenosis | n / % | 45 | 1.5% | - | - |  
Tetralogy of Fallot | n / % | 43 | 1.4% | - | - |  
Left-sided superior vena cava | n / % | 23 | 0.8% | - | - |  

Table of baseline characteristics from the English registry compared to a 2014 meta-analysis of subaortic stenosis

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*Meta-analysis by Etnel et al., European Journal of Cardio-Thoracic Surgery (2014) 1–9*
Concurrent surgical procedures at initial and redo surgery: $n$ (left), $\%$ (right)
Results

- 1,632 procedures were carried out in 1,349 patients.
- Median follow-up period of 9.4[0–18] years.
- Overall survival:
  - 30 day: 99.3% (98.7–99.6)
  - 1 year: 98.8% (98.1–99.3)
  - 10 years: 85.2% (82.5–87.6)
  - 15 years: 79.5% (75.7–82.9)
- 15-year survival in adults was high at 94.9% (88.9–97.7), but in infants this was 58.7% (39.8–73.4).
- Female sex (p = 0.03) and younger age (p<0.0001) were independent predictors of re-intervention; the type of surgery and common associated lesions were not.
Study limitations

• Retrospective study design.

• Administrative NHS database with limited clinical information:
  – We have to rely on accurate coding;
  – Lacks the granularity seen with prospectively collated single-centre data.

• ICD-10 and OPCS-4 codes have limitations:
  – No information about echocardiographic or catheter data;
  – Cannot distinguish between discrete and diffuse SubAS.

• Patients with SubAS not admitted to hospital during the study period are not captured.
Conclusions

• SubAS patients have benefited significantly from advances in surgery over the last several decades.

• However, reoperation rates remain high in this contemporary cohort.

• This is especially true of patients repaired early in life, who are likely to present with more severe forms of the disease.

• Further work is needed to optimise the outcome of this cohort of patients.