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EDITORIAL
Congenital heart disease

Subvalvular aortic stenosis in adulthood: stable, manageable, but never simple

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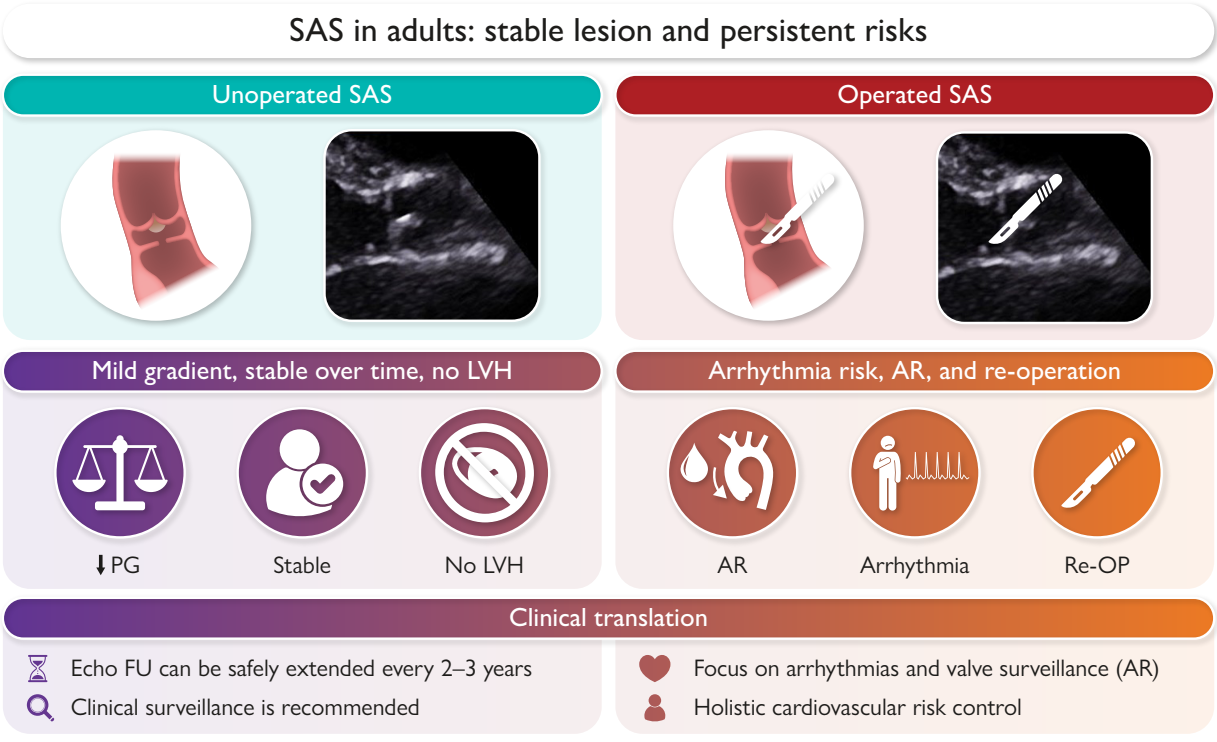
This editorial refers to 'Subvalvular aortic stenosis in adults: clinical course and long-term outcomes', by A.R. de Keijzer *et al.*, https://doi.org/10.1093/eurheartj/ehaf1078.

Graphical Abstract



Graphical summary of the study by de Keijzer *et al. European Heart Journal*, showing that in adulthood, SAS is characterized by haemodynamic stability—reflected by minimal gradient progression and absence of LVH—but remains associated with meaningful morbidity. Unoperated SAS generally shows mild, stable gradients and no LVH, whereas operated patients, despite similar haemodynamics, experience a persistent risk of arrhythmia, AR, and reoperation Re-OP. Clinical translation: echocardiographic follow-up can safely be extended to every 2–3 years in stable, unoperated adults, while continued clinical surveillance is warranted after surgical repair, with emphasis on arrhythmia monitoring, AR progression, and comprehensive cardiovascular risk management

AR, aortic regurgitation; **FU**, follow-up; **LVH**, left ventricular hypertrophy; **PG**, pressure gradient; **Re-OP**, reoperation; **SAS**, subvalvular aortic stenosis

Sabatino J, *et al. European Heart Journal.*

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The evolution of congenital heart disease (CHD) management from paediatric repair to lifelong care has reshaped modern cardiology.[1,2] As the adult congenital heart disease (ACHD) population expands, clinicians face the dual challenge of managing congenital residua alongside acquired cardiovascular disease.[3,4] Yet major evidence gaps persist, as most guidelines extrapolate from acquired cohorts rather than ACHD-specific data. A new generation of population-based studies is now redefining risk stratification and follow-up strategies tailored to congenital phenotypes.[5,6]

Among the lesions that have eluded precise characterization, subvalvular aortic stenosis (SAS) stands out. Once considered an exclusively paediatric condition, SAS is increasingly encountered in adult congenital practice.[7,8] Despite decades of experience with valvular and supravalvular obstruction, the adult natural history of SAS has remained unclear, particularly for residual or unoperated forms. In this issue of the *European Heart Journal*, de Keijzer and colleagues provide the most comprehensive analysis to date on SAS, drawing on nearly two decades of nationwide data from the Dutch CONCOR registry.[9] Their findings close a critical evidence gap, clarify prognosis, and suggest a reassessment of how these patients are monitored throughout adulthood.

A nationwide, long-term perspective

Subvalvular aortic stenosis, typically involving a discrete fibromuscular or fibrous membrane beneath the aortic valve, accounts for ∼8%–20% of left ventricular outflow tract (LVOT) obstructions.[7,8] While morphological expression ranges from thin ridges to tunnel-like narrowing, the haemodynamic consequence is fixed obstruction with potential for left ventricular hypertrophy (LVH) and secondary aortic regurgitation (AR). Most patients undergo surgical resection in childhood, yet the trajectory of residual or newly diagnosed adult SAS has remained anecdotal.[10]

Leveraging the CONCOR registry, which prospectively enrols adults with CHD from all Dutch centres, the authors identified 312 adults with SAS (mean age 26 years, 52% male) followed for a median of 16 years—the longest observation ever reported for this type of CHD. Two-thirds had prior surgical repair, allowing meaningful comparison between operated and unoperated cohorts within a consistent national care framework. Exclusion of hypertrophic cardiomyopathy and univentricular anatomies ensured a well-defined and representative SAS population.

A stable lesion in adulthood

The study's most striking message is that SAS in adults is remarkably stable. Across thousands of serial echocardiograms, progression of LVOT velocity was modest (0.1 m/s in the first decade, 0.3 m/s thereafter), with no patient showing rapid acceleration (≥0.3 m/s per year). Ventricular wall thickness remained normal and no frank LVH developed. These data convincingly argue against progressive remodelling once growth is complete, at least in mild and moderate forms. Clinically, this challenges the prevailing routine of frequent echocardiographic surveillance, often borrowed from paediatric or valvular paradigms. The authors suggest that less frequent imaging may be adequate for asymptomatic adults without AR, provided structured ACHD follow-up is maintained. This is a powerful translational message: evidence-based de-escalation of surveillance can reduce patient burden and system cost while preserving safety.

Survival and morbidity: stable anatomy, persistent risk

Overall survival was excellent, mirroring the matched general population for unoperated patients, but lower in those with prior repair (15 year survival ∼90% vs 99%). The difference is likely to reflect baseline severity rather than surgical harm. Importantly, the operated cohort remains vulnerable to late morbidity, underscoring that surgical 'cure' does not equate to normalized risk. The 15 year cumulative incidence of (re-)operation for AR was 7.6%, while arrhythmias were common, affecting two-thirds of patients during follow-up. Pacemaker implantation, heart failure admissions, and other interventions contributed substantially to the lifetime clinical course. Thus, adult SAS may be haemodynamically quiet yet clinically active, its late complications driven more by post-operative scarring and electrical remodelling than by LVOT gradient progression itself.

Sex and associated lesions

A male predominance (∼2:1) was confirmed, but no significant sex differences were seen in survival or event rates after adulthood. Similarly, the presence of additional congenital lesions, in ∼30% of cases, did not materially affect long-term outcomes. These findings reinforce the concept that, once somatic growth has ended, disease behaviour depends primarily on individual baseline severity rather than sex or associated anatomy.

Implications for clinical practice

For most adults with mild, unoperated SAS, the message is reassuring: the lesion is stable, progression slow, and prognosis excellent. Echocardiographic follow-up can therefore be safely extended to every 2 or 3 years, allowing clinicians to redirect attention towards what truly matters: arrhythmia surveillance, AR monitoring, and general cardiovascular risk control. For patients with previous repair, vigilance remains essential. Lower survival despite stable haemodynamics suggests the importance of long-term rhythm monitoring, blood pressure management, and early identification of AR (see *Graphical Abstract*). Post-operative follow-up may benefit from refocusing imaging goals, from residual gradient quantification to assessment of aortic valve integrity and LV geometry.

This evidence directly informs the 2020 ESC Adult CHD Guidelines, which provide only limited detail on adult SAS.[11] The study justifies a more relaxed imaging cadence for mild, stable disease and supports individualized risk-based intervals, aligning SAS management with other stable congenital defects such as small residual ventricular septal defects (VSDs) or mild supravalvular stenosis. For patients with prior repair or relevant AR, annual clinical review remains prudent.

How does SAS stabilize after growth?

The biological mechanisms underlying this apparent stability remain unclear but intriguing. Once somatic growth ceases, the fibromuscular membrane may lose a key proliferative stimulus, or the myocardium may adapt through geometric and cellular remodelling that mitigates shear stress.[12] Alternatively, developmental 'programmes' that drive early LVOT remodelling may simply switch off at the end of physiological growth, leaving behind a fixed anatomical substrate with relatively

stable haemodynamic consequences. Addressing these questions will require going beyond imaging and clinical observation into the study of developmental biology. SAS arises in a cardiac region formed by complex interactions between endocardial cushions, neural crest-derived cells, and flow-dependent signalling pathways during valvulo-arterial morphogenesis.[13] Dissecting how perturbations in these pathways translate into discrete subaortic membranes, tunnel-like narrowing, or associated valvular abnormalities will demand integrated studies that link embryology, genomics, and mechanobiology. In this context, human induced pluripotent stem cell-derived cardiac organoids offer an attractive experimental model system.[14] Three-dimensional organoid models that recapitulate key aspects of LVOT and aortic root development could be used to study how specific genetic variants, signalling cues, or shear stress patterns drive subvalvular fibromuscular proliferation, and how these responses change over developmental time. This experimental platform may also offer unique mechanistic insight into how SAS is dynamic in childhood yet quiescent in adulthood. Ultimately, such developmental and organoid-based approaches could fill the gap between genotype, morphogenesis, and adult phenotype, opening the door to earlier risk stratification and, one day, personalized medicine strategies.

Beyond the gradient: toward holistic care

The CONCOR findings also symbolize the maturation of adult congenital cardiology itself. National registry infrastructure, long-term data harmonization, and multidisciplinary collaboration have transformed isolated institutional series into population-level science. This shift allows clinicians to move beyond anatomy and gradient towards lifelong cardiovascular management in anatomically unique hearts. Adults with SAS—even those deemed 'stable'—require integrated care addressing rhythm surveillance, reproductive counselling, and psychosocial well-being. The study empowers clinicians to personalize this care with confidence, basing decisions on evidence rather than extrapolation.

A dual message: reassurance and vigilance

For clinicians, the takeaway is reassuring, though it comes with a cautionary tale. In adults, SAS demonstrates minimal progression and near-normal life expectancy, yet post-operative patients remain at higher risk, and arrhythmias or heart failure can emerge even in apparently stable anatomy. For guideline committees, these data offer a rare opportunity to refine surveillance protocols based on empirical, long-term evidence.

Future studies should refine risk prediction, distinguishing which adults truly need annual imaging from those safely reviewed every few years, and explore the interplay between surgical history, electrical remodelling, and late mortality. Integration of wearable rhythm monitoring, longitudinal tissue characterization by magnetic resonance imaging (MRI), and patient-reported outcomes will further redefine what 'stability' means in congenital cardiology.

Conclusions

The work by de Keijzer *et al*. sheds new light on subvalvular aortic stenosis in adulthood. It reveals a condition that is not relentlessly progressive but predictably stable, where the greatest risks arise from sequelae rather than obstruction itself. As the adult congenital population grows, evidence from nationwide registries like CONCOR will be indispensable to guide personalized, resource-conscious care. SAS may indeed be a stable lesion—but, as this study elegantly reminds us—stability is never the same as simplicity.

Declarations

Disclosure of Interest

The authors declare no disclosure of interest for this contribution.

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