JAMA Surgery | Review

Pectus Arcuatum Definitions, Diagnostics, and Surgical Guidelines
A Systematic Review

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IMPORTANCE Pectus arcuatum is a rare anterior chest wall deformity that is often confused with excavatum, carinatum, or a mixed deformity. A complete understanding of its clinical and physiological basis will improve diagnostic accuracy and allow for standardized management.

OBJECTIVES To gain a comprehensive understanding of pectus arcuatum, focusing on its nomenclatures, epidemiology, classification, pathophysiology, preoperative presentation and assessments, management options, and surgical outcomes.

EVIDENCE REVIEW PubMed, Google Scholar, and subsequent reference searches were used to extract original studies addressing pectus arcuatum fully or partially. The searches were performed on February 2, 2025. Two independent reviewers screened articles based on preestablished inclusion and exclusion criteria. Quality assessment was performed, but given the rarity of arcuatum and the limited, heterogeneous nature of available literature, none of the studies were excluded based on the level of evidence or quality assessments. Using a standardized data extraction form, results were synthesized and presented as a review with recommendations to guide clinical decision-making.

FINDINGS A total of 65 studies were included in the final review. Of those, 37 (56.9%) were cohort studies, 18 (27.7%) were case reports, 8 (12.3%) were case series, and 2 (3.1%) were technical reports. A total of 455 cases of pectus arcuatum were represented. Heterogeneity was found in nomenclature, epidemiology, associated conditions, assessments, management, and outcomes. An open surgical approach and, more recently, hybrid open and minimally invasive technique are reported for surgical repair. Postoperative outcomes were good in most surgical series, with no significant difference when compared with the repair of other pectus deformities.

CONCLUSIONS AND RELEVANCE Results of this systematic review reveal that pectus arcuatum is a rare variant of pectus deformities that is distinct from carinatum and excavatum. Unified terminology and standardized diagnostic criteria are important to facilitate proper management. Minimally invasive hybrid approaches continue to refine outcomes, balancing invasiveness with efficacy.

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Supplemental content

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Pectus arcuatum is a unique anterior chest wall deformity involving premature fusion of the sternal ossification centers and obliteration of the manubriosternal joint. It may be misdiagnosed as excavatum, carinatum, or mixed deformity. The literature shows significant variation in clinical aspects and terminology. Difficulties in obtaining clear information and misunderstanding of the deformity have hindered accurate assessment and standardized management. This systematic review aims to provide a comprehensive summary of the current literature to guide diagnostic and treatment recommendations.

Methods

Study Design

This systematic review provides a comprehensive summary of arcuatum, focusing on its nomenclature, epidemiology, classification, pathophysiology, preoperative presentation and assessments, management, and outcomes. It followed the Preferred Reporting Items for Systematic Reviews and Meta-analyses (PRISMA) reporting guidelines; the protocol is available on request.

Search Strategy

A structured search was conducted in PubMed and Google Scholar databases using MeSH terms and free-text keywords. PubMed used the following search terms: ("Pectus Carinatum"[Mesh]) OR (Currarino-Silverman syndrome) OR (Type II pectus carinatum) OR (Pouter pigeon chest) OR ("Horns of Steer") OR (Chondromanubrial deformity) OR (arcuatum). Google Scholar used the following search terms: ("Pectus Carinatum") OR (Currarino-Silverman syndrome) OR (Type II pectus carinatum) OR (Pouter pigeon chest) OR ("Horns of Steer") OR (Chondromanubrial deformity) OR (arcuatum). No time limits were set, and the search was performed on February 2, 2025.

Screening and Selection

Initial search results were deduplicated and screened by title and abstract using an artificial intelligence–powered tool. Ambiguous or relevant papers underwent full-text screening. Additional articles were identified through citation tracking. Two reviewers (R.M.Z., A.E.K.) independently screened studies. Disagreements were resolved by consensus with an expert (D.E.J.).

Inclusion Criteria

- Studies specifically addressing arcuatum (fully or as part of a mixed cohort).
- English-language publications.
- Technical papers and original studies (randomized clinical trials, cohort, case-control, series, reports).

Exclusion Criteria

- Nonhuman studies.
- Full article text not available.
- Reports of superior sternal protrusion or mixed deformities without clear evidence of arcuatum.
- Studies on various pectus deformities without stratified results by type.

Data Extraction

A single expert reviewer (R.M.Z.) extracted data using a standardized form (eTable 1 in the Supplement) to ensure accuracy and capture of nuances specific to arcuatum.

Quality Assessment and Data Synthesis

Study quality was assessed using ROBINS-I for observational studies and JBI tools for case series/reports (eTable 2 in the Supplement). Given the rarity and heterogeneity of arcuatum studies, none were excluded based on level of evidence or quality. All relevant studies were included for comprehensive coverage. No meta-analysis was conducted. Outcomes were synthesized descriptively where available.

Results

From the database screening, 971 articles were identified. After removing 46 duplicates, 925 papers underwent title and abstract screening. Of these, 260 studies were selected for full-text review. Applying eligibility criteria and adding 8 papers from reference screening, 65 papers were included. Of those, 37 (56.9%)[1-37] were cohort studies, 18 (27.7%)[38-55] were case reports, 8 (12.3%)[56-63] were case series, and 2 (3.1%)[64,65] were technical reports. Cohen κ of 0.7909 indicates substantial reviewer agreement. A total of 455 arcuatum cases were represented. Study characteristics are summarized in eTable 3 in the Supplement; the PRISMA flow diagram appears in the eFigure in the Supplement.

Nomenclatures

Arcuatum (**Figure 1**) has been referred to by various names as follows: (1) Currarino-Silverman syndrome,[56] (2) angulated synostosis,[1] (3) arcuate deformity,[2] (4) chondromanubrial deformity,[66] (5) manubriosternal prominence,[3] (6) type II pectus carinatum,[4,5] (7) horns of steer deformity,[57] (8) pouter pigeon breast deformity,[6,38] (9) pectus carinatum superior,[7] and (10) Wenlin chest.[39]

Key Points

Question What are the standardized definitions, diagnostics, and surgical guidelines for pectus arcuatum?

Findings In this systematic review including 65 studies, preoperative computed tomography/magnetic resonance imaging and echocardiography were recommended to assess cardiopulmonary function and screen for congenital heart disease. Symptomatic patients should be considered for surgery, with expected improvement.

Meaning Results suggest that surgical correction relied on wedge osteotomy, with titanium plating and bar placement being key for stabilization and lower cartilage remodeling. Hybrid techniques continue to evolve, aiming to balance invasiveness and efficacy.

Epidemiology

The prevalence and demographics of arcuatum remain unclear in part due to an underreporting and misdiagnosis of these deformities. Arcuatum is commonly considered a rare pectus variant, accounting for 1% or fewer cases.[8,58] It is likely more prevalent. In a 10-year study, Kelly et al[9] found 7% of patients (31 patients) had arcuatum. Haje et al[10] reported 5% among 4012 patients over 32 years.

Our review identified 455 cases: 15% female, 22% male, 63% unreported (eTable 3 in the Supplement). Lacquet et al[11] reported equal sex distribution. Zeineddine et al[12] noted slight male predominance (11 males vs 9 females).

A recent study (2017-2021)[13] of 1520 children with anterior chest wall deformities found 17 arcuatum cases: 6 Hispanic, 10 non-Hispanic White, and 1 unspecified race and ethnicity.

Classification

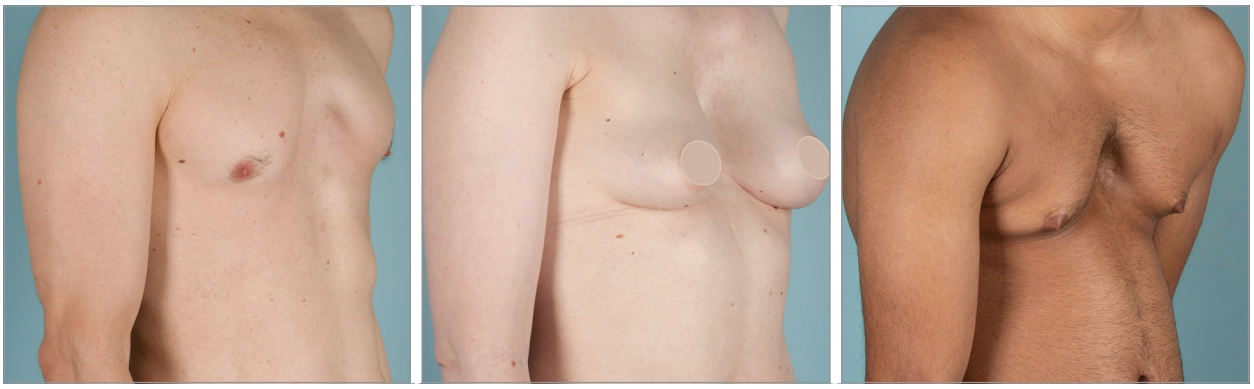
Despite the description by Currarino-Silverman of the hallmarks of arcuatum in 1958,[56] its unique combination of features, some overlapping with other deformities, has led to the deformity being misdiagnosed as other pectus variants.

Some viewed arcuatum as a type of protrusion deformity or carinatum variant. In 1952, Lester divided protrusion deformities into midline (sternum/cartilage) and lateral anterior chest wall types, further separating midline into upper arcuate and lower oblique forms.[2,14] In 1957, Lester categorized pectus into excavatum, carinatum, or mixed types, proposing a shared etiologic basis.[14] In 1958, Brodkin identified 2 protrusion types: upper chondromanubrial and lower chondrogladiolar prominence, the former involving manubrium and adjacent first and second cartilages with a depressed gladiolus.[59] Robiscek et al[5] divided carinatum into 3 types, with type II (pouter pigeon breast) described as a variant featuring chondromanubrial prominence, protrusion of the first 2 cartilages, a posteriorly arched sternal body, and an anteriorly pointing xiphoid tip. In 1999, Haje et al[7] categorized pectus into 5 types based on sternal protrusion/depression localization, referring to arcuatum as *pectus carinatum superior*.

Given the lower degree of depression seen in arcuatum, some consider it an excavatum variant.[8,57] Others suggest arcuatum as a mixed deformity.[15] In 1991, Saxena and Willital[15] classified chest wall deformities into 11 types with arcuatum (type 9) being a transitional form: funnel chest (4 types), pigeon chest (4 types), funnel-pigeon combinations (type 9), thoracic wall aplasia (type 10), and cleft sternum (type 11).

Figure 1. Physical Features of Pectus Arcuatum

A Male with typical physical features of arcuatum B Female with typical physical features of arcuatum C Associated kyphosis



A-C, Typical physical features of pectus arcuatum: manubriosternal prominence, lower sternal depression, and abnormally inserted upper ribs forming the characteristic "horns of a steer" appearance.

Acastello[67] classified chest wall deformities by defect origin: cartilaginous (type 1), costal (type 2), chondrocostal (type 3), sternal (type 4), and clavicle-scapular (type 5), attributing arcuatum to cartilaginous type 2 or superior carinatum.[16]

Recently, surgeons have stressed on distinguishing arcuatum from other pectus deformities, emphasizing that it is not a mixture of these conditions.[12,17,18]

Pathophysiology

Arcuatum was recognized as a congenital deformity but often unnoticed until adolescence.[14,59] Brodkin proposed his diaphragmatic theory in 1949[66] with reinforcement in 1957,[59] suggesting unbalanced contractions of a malformed diaphragm (*V*-shaped fibrotendinous structure with thin anterior and hypertrophied lateral segments) drive the deformity. These forces push the sternum and cartilage forward, causing upper protrusion and lower retraction during inspiration. As the thorax elongates, upper protrusion persists while the lower gladiolus flattens. Limited evidence without control groups hinders confirmation of observed patterns as unique to arcuatum.[59]

In 1952 and 1957,[2,14] Lester combined published observations[66] with intraoperative findings, suggesting disproportionate second to fourth rib overgrowth and diaphragm shape drives the deformity. Rib overgrowth creates force and buckles at weak points, typically the costochondral junction, pushing the sternum forward. If the diaphragm fails to adjust, the xiphoid restrains it, causing arcuate sternal protrusion.

Premature sternal ossification centers fusion and manubriosternal joint obliteration were coined by Currarino-Silverman in 1958[56] and supported by others.[1,7] Normally, ossification begins at 5 to 6 months in fetal development, with segments fusing gradually. Complete sternal fusion is rare, and manubriosternal fusion occurs in only 10% of adults. However, arcuatum patients uniformly exhibit early manubriosternal fusion, confirmed radiologically.[7,12,60] Fokin[1] compared premature sternal fusion radiograms with earlier ones, found abnormally narrowed sternal sutures, supporting premature obliteration over failed segmentation. Causes include undeveloped fibrous lamina or pathological joint degeneration.[56] Ravitch supported Currarino-Silverman, emphasizing rib overgrowth as primary drivers of arcuatum rather than sternal abnormalities. He dismissed the diaphragmatic theory, arguing that the diaphragm only accentuates the deformity. He suggested rib lateral incurvature makes the sternum appear prominent and compensates for the latent depressed area,[6] but in arcuatum, the ribs are bulky and override the sternum rather than curve inward.[12] Fokin also identified second to fifth cartilage overgrowth with hyaline cartilage degeneration, atypical fibrils, reduced chondrocytes, and thin periosteum.[1]

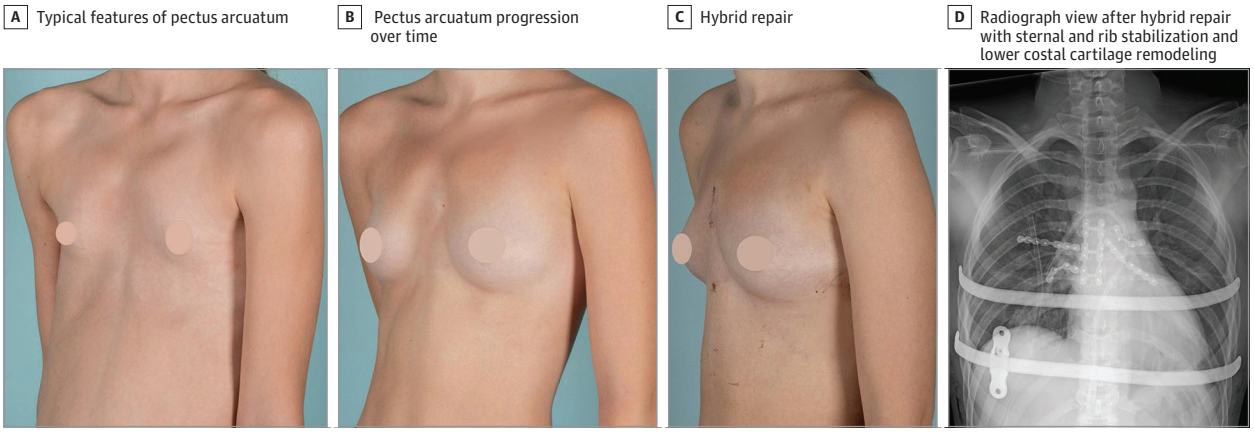
Abnormal differentiation of anterior mesenchymal cells during cardiogenesis may disrupt endocardial cushions, sternum, and aortic derivatives, explaining arcuatum's link to congenital heart diseases (CHDs) and supporting a potential genetic component.[68]

Preoperative Presentation and Assessment

History

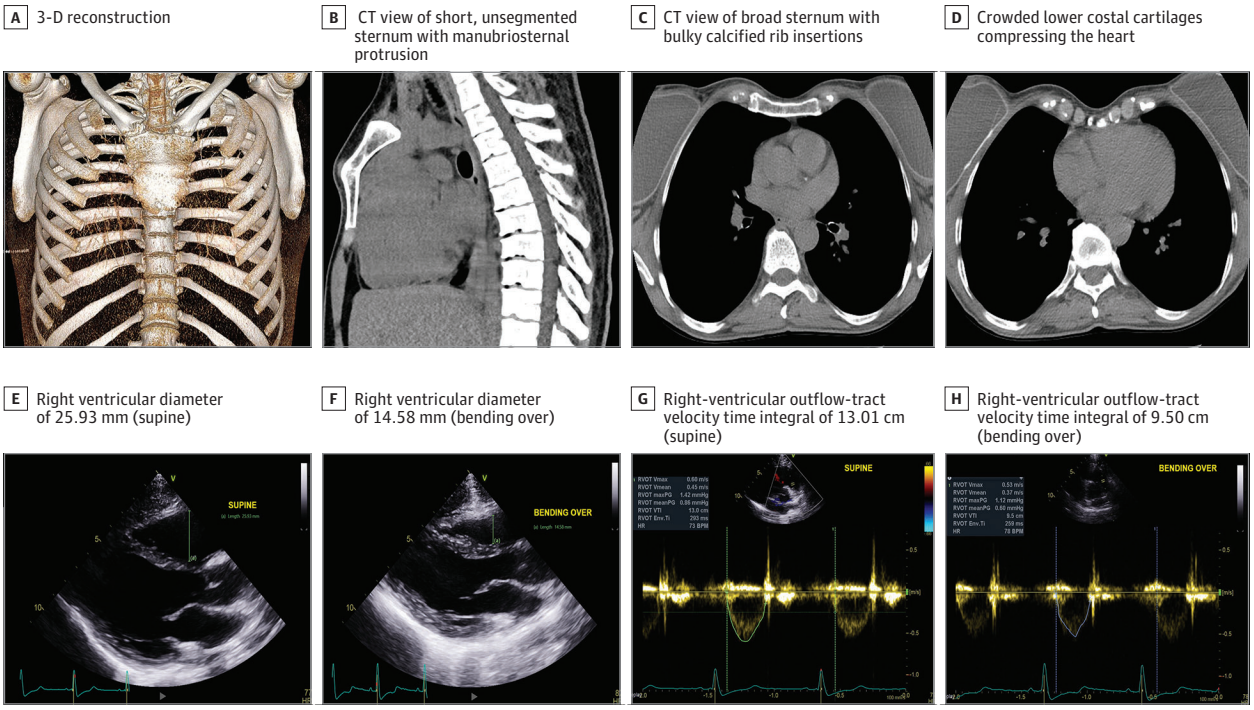
- Congenital: typically noticeable before age 3 years, often within the first year of life.[1,18,38,56,59]
- Progression: Fokin observed progression in 50% of patients (n = 18), with a 2° per year nonlinear progression rate uncorrelated with lower sternal depression.[1] Currarino-Silverman reported 1 case with straight sternum at 1 year, featuring narrowed, irregular sutures, became angulated by age 4 years, and another with arcuatum evident at 8 months, worsening over time.[56] Multiple reports supports progression (**Figure 2**A and B).[40-42]
- Psychosocial implications: the psychosocial effects are significant. Patients often avoid chest-revealing activities/clothing, show abnormal posture, low self-esteem, anxiety, or depression.[1,12,16,59,64] Fokin noted psychological effects begin around age 7 years and peak by 10 years,[1] although cosmetic concerns persist into adulthood.[12] Many prefer surgical scars over the deformity, sometimes viewing them as symbols of victimhood, reflecting the profound psychological impact.[1]
- Cardiovascular symptoms: chest pain and exercise intolerance are most common.[1,12,19,61] Cardiac compression, displacement, and pulmonary artery compression can result from inward lower sternum and cartilages angulation (**Figure 3**D).[12,38,43]

Figure 2. Patient With Progressive Features of Pectus Arcuatum: Before and After Hybrid Repair



Pectus arcuatum evident from a young age (A), which progressed over time (B). The patient subsequently underwent hybrid repair (C), during which her sternum was stabilized using an H plate after a single wedge osteotomy, her abnormal ribs were reapproximated and rigidly fixed bilaterally, and 2 bars were placed to correct her lower sternal depression. One bar was positioned at a low point below the sternal end to support lower costal cartilage remodeling and relieve cardiac compression (D).

Figure 3. Preoperative Assessments



A 3-dimensional (3-D) reconstruction (A) and computed tomography (CT) views of a short, unsegmented (B), broad sternum with bulky and calcified rib insertions (C) and crowded lower costal cartilages compressing the heart (D). Positional transthoracic echocardiogram demonstrates right ventricular compression at the lower costal cartilages, with the right ventricular diameter decreasing from 25.93 mm (supine) (E) to 14.58 mm (bending over) (F) and the right ventricular outflow tract velocity-time integral decreasing from 13.01 cm (supine) (G) to 9.50 cm (bending over) (H).

- Pulmonary issues: pulmonary symptoms are debated. Some link them to decreased respiratory excursion and increased anterior-posterior diameter, leading to infections.[12,19] Others consider them uncommon and milder than in excavatum.[1] Postoperative improvement and reduced asthma have been reported.[20] Pulmonary evaluation may help, as cases of diaphragmatic dysfunction, restrictive/obstructive diseases, and recurrent infections exist.[1,20,62]
- Family history: many patients with pectus arcuatum have relatives with the condition. Chidambaram reported arcuatum in 2 siblings and their mother.[60] Fokin found 4 of 11 patients had close relatives with chest deformities, 2 with confirmed parental

sternal synostosis.[1] Abdellaoui et al[18] reported 32% (n = 34) with skeletal malformation family history. Zeineddine et al[12] noted 7 of 16 patients had a family history of pectus, none being arcuatum. Currarino-Silverman included 3 cases, 1 with an affected maternal aunt, and frequent manubriosternal synchondrosis in parents.[56] Inheritance appears polygenic or multifactorial. Some reported shared genetic links across anterior chest deformities.[14,62,69]

- CHD: arcuatum has been associated with various CHD (Table).[1,11,12,16-18,21,40,42,44-48,56,58,60,62,64] Chidambaram et al[60] noted mitral valve prolapse with mild regurgitation in 1 sibling and their mother.
- Syndromic associations: arcuatum may be isolated or associated with connective tissue diseases, spinal deformities, and syndromes. Lacquet et al[11] reported 3 of 37 hemiarcuatum cases over 25 years, associated with contralateral Poland syndrome. Kuzmichev et al[64] noted occasional Poland syndrome. Zeineddine et al[12] reported 1 of 20 patients with Klinefelter syndrome. Noonan syndrome has also been reported.[18,47,48] The Table summarizes all anomalies.[1,11,12,16-18,21,40,42,44-48,56,58,60,62,64]

Physical Examination

- Chest morphology: upper sternal convexity and lower concavity with prominent rib insertions create the classic horn of steer appearance (Figure 1A-C).
- Functional murmurs: Fokin reported functional murmurs in 7 of 18 patients, likely caused by right ventricular outflow tract obstruction.[1]
- Associated anomalies: evaluation for evidence of connective tissue disorders or other musculoskeletal abnormalities.

Imaging

Radiographs | Chest radiograph and computed tomography (CT) are essential for preoperative assessment; 3-dimensional CT or magnetic resonance imaging are considered best.[16,18] Findings include upper sternal protrusion, lower depression, and abnormal costosternal junctions. The sternum is short, broad, and nonsegmented (1-2 segments) (Figure 3A-C).[6,7,12,18,22,56] Manubriosternal fusion has been reported,[7] with a reduced angle of Louis (normal: 175°-145°), increasing sagittal chest diameter.[1,6,18,56] Acute proximal third sternal angulation is common in arcuatum, unlike the gradual curvature in carinatum.[22] The manubrial orientation also differs from excavatum, where it is more vertical.[23] A *Z*-shaped sternum, or angulated synostosis, appears on lateral views.[1,6]

The xiphoid-vertebral body distance is often normal, yielding a normal Haller Index.[12,14,24] Lower sternum depression occurs in at least one-third of cases.[1] The lower sternal end tends to be vertical, unlike excavatum (posterior) and carinatum (anterior).[22] Less commonly, the xiphoid may be asymmetric, bifid, or absent.[6,16,63]

The deformity is usually symmetric.[1] Costosternal insertions are often bulky and heavily calcified (hyperostosis), primarily affecting second to fourth/fifth ribs.[1,12,25,26,63] Lower cartilages are often longitudinally clustered at their sternal insertion,[12,58] possibly causing right ventricular compression.[12,43] The inward buckling and compression from these lower cartilages can be position-dependent and may be missed on supine imaging (Figure 3D-H).[12]

Table. Details of the Articles Reporting Associated Congenital Heart Diseases Among Other Anomalies

Source	Cases with pectus arcuatum, No.	Associated anomalies
Associated congenital heart diseases		
Currarino and Silverman,[56] 1958	3	(1/3) Large PDA, large VSD, pulmonary artery and arteriolar stenosis
Briinner,[44] 1961	1	ToF and PDA
Kaplan et al,[45] 1968	1	Pulmonic stenosis
Welch and Vos,[62] 1973	2	(1/2) Double aortic arch and PDA
Chidambaram and Mehta,[60] 1992	5	(1/5) Parachute mitral valve with mild mitral stenosis and regurgitation, (1/5) endocardial cushion type of atrioventricular valve with mild to moderate MR, (2/5) MVP with mild MR, (1/5) aortic coarctation, (1/5) ostium secundum type of ASD that closed spontaneously
Mehta,[46] 1996	1	Aortic coarctation and parachute mitral valve
Fokin,[1] 2000	18	(2/18) VSD, (1/18) ToF
Kuzmichev et al,[64] 2016	NA	Unspecified CHD
Hysi et al,[21] 2015	3	(1/3) ASD, (1/3) ToF, recurrent VSD and pulmonary and aortic stenosis
Gritsiuta et al,[16] 2021	4	(1/4) MR and tricuspid regurgitation, (1/4) MVP (3 mm) and VSD (3 mm), (1/4) MVP (5 mm)
Abdellaoui et al,[18] 2023	34	(2/34) Unspecified CHD
St-Louis et al,[17] 2024	5	(1/5) Mild anterior mitral valve leaflet thickening
Other associated anomalies		
Currarino and Silverman,[56] 1958	3	2 Kyphosis, 1 kyphoscoliosis, case 1: mild congenital micrognathia, bilateral clubfoot, inguinal hernia, and cryptorchidism. Case 2: underdeveloped physical growth, craniofacial asymmetry, occipital and frontal flattening, microphthalmia, cataract, incontinentia pigmenti, aberrant subclavian artery causing dysphagia
Kaplan et al,[45] 1968	1	NS
Shamberger and Welch,[58] 1988	5	1 Bilateral clubfeet
Lacquet et al,[11] 1998	37	3 Poland syndrome
Fokin,[1] 2000	18	1 Ossification of T3-T4 disk
Kuzmichev et al,[64] 2016	NA	Poland syndrome
Alkhunaizi et al,[47] 2018	1	NS
Sahu et al,[48] 2020	1	NS
Gritsiuta et al,[16] 2021	4	2 Kyphosis, 1 kyphoscoliosis
Wang et al,[40] 2022	1	Poland syndrome
Wang et al,[42] 2022	1	Scoliosis
Abdellaoui et al,[18] 2023	34	4 NS, 3 scoliosis, 1 cervical rib, 1 cervical-dorsal vertebral malformation, 1 Baraitser-Winter syndrome, diaphragmatic hernia
Zeineddine et al,[12] 2024	20	1 Klinefelter syndrome, 6 scoliosis

Abbreviations: ASD, atrial septal defect; CHD, congenital heart diseases; MR, mitral regurgitation; MVP, mitral valve prolapse; NA, not available; NS, Noonan syndrome; PDA, patent ductus arteriosus; ToF, tetralogy of Fallot; VSD, ventricular septal defect.

Echocardiography | Echocardiography is recommended preoperatively due to the high association between arcuatum and CHD.[1] Positional echocardiography is particularly effective in detecting transient cardiac compression during positional changes, often missed in standard examinations (Figure 3E-H).[12]

Other Tests | Cardiopulmonary exercise testing (CPET) has been used in patients with exercise limitation. Zeineddine et al[12] reported decreased maximal oxygen consumption (VO_2; median, 67.0%; IQR, 57.3%-69.0% of predicted) and mild restrictive patterns in 38.5% on pulmonary testing. González-Barba et al[24] found no CPET abnormalities in 3 patients.

Management

Noninvasive Approach

As a rule, bracing is largely ineffective in arcuatum due to early sternal rigidity and is not recommended.[9,18,27-30] Haje et al,[10] however, reported outcomes for his bracing protocol, which included individualized orthosis, prolonged treatment, exercise to balance internal and external pressures, close follow-up, and expert pressure adjustments. In his 32-year study of 69 children and 4 adults with arcuatum, 7.3% of children had good-to-excellent improvement and 21.7% moderate improvement.[10] In adults, only 1 patient noted some moderate improvement.[10]

Suggested Surgical Indications

Cosmetic dissatisfaction is the most reported surgical indication.[16,17,24,49,59,64] Others emphasized symptoms and cardiopulmonary compromise as indication for surgery.[9,12,20] Fokin provided recommendations based on disease progression, syndromic associations, cardiopulmonary findings, and angle of Louis:

- Mild deformities (angle >130°): annual follow-up.
- Moderate deformities (angle 130°-115°): surgery, especially with lower depression.
- Severe deformities (angle <115°): surgery.

Surgical Approach

The evolution of arcuatum surgical techniques reflects ongoing refinement. Ravitch introduced the first operative approach in 1952, involving extensive cartilage resection and sternal osteotomies.[6] Over decades, surgeons modified and supported this open technique.[9] In 1957, Chin proposed xiphosternopexy, reattaching the xiphoid at a higher level to reposition the sternum over time, though it was not applied to arcuatum.[3] Brodkin later referenced it for 2 cases of unilateral chondromanubrial deformities.[15,59] Although Chin's xiphosternopexy supported Brodkin's diaphragmatic theory, concerns remain. Brodkin described chondromanubrial deformity as an upper protrusion with a flat or depressed lower part featuring the Harrison groove, sometimes unilateral. This describes a carinatum variant, not arcuatum. He later noted surgery was ineffective for non-unilateral cases, which likely represent true arcuatum.[59] Howard was successful with Chin's method only after modifications with cartilage and sternal resections.[30]

Advancements in surgical expertise have led to a shift toward less invasive procedures.[12,17,29] Since Onen introduced the hybrid technique in 2016,[31] it has shown good outcomes (Figure 2 and **Figure 4**) with low complications.[12,17,18,24,29,31,32,50] eTable 4 in the Supplement outlines the chronological progression of techniques.

Age recommendations for surgery remain controversial. Some advocate waiting for rib growth completion,[64] whereas others prefer early adolescence when cartilage is most flexible and surgery less demanding.[11,20] Challenges with younger patients include postoperative noncompliance and thoracic dystrophy,[11,16] whereas older patients face greater surgical difficulty due to sternum rigidity.[11] For patients with connective tissue disorders, surgery is advised after growth completion due to high regression risk.[11]

For patients with concurrent cardiac abnormalities, whether to perform surgery concurrently or in stages remains debated. Hysi et al[21] recommend Ravitch for optimal exposure and postoperative management. The hybrid approach is an option but may risk reduced cardiac compression efficacy and increased midsternotomy dehiscence. If intrathoracic bars are placed, pericardial closure or a barrier is required to prevent cardiac adhesions that may cause injury during bar removal.[21,70]

Postoperative Complications and Outcomes

Few complications are reported in the literature,[1,9,17,24] likely due to the lack of stratified, dedicated studies. For the open approach, Ravitch[38] reported in his first case of right pleural effusion requiring thoracentesis. Kelly et al[9] reported in their 10-year experience 1 wound infection managed with antibiotics. For the hybrid approach, Zeineddine et al[12] reported few complications, the most significant being 1 hemothorax requiring pigtail insertion and transfusion and the other complications being 3 pleural effusions requiring thoracentesis.

Recovery was satisfactory and comparable to other pectus deformities.[1,12,25,64] Most had good cosmetic outcomes[1,11,12,24,38]; however, residual prominence or asymmetry due to inadequate resection was reported.[11] Long-term cardiopulmonary improvement was subjectively validated by patients.[1,12]

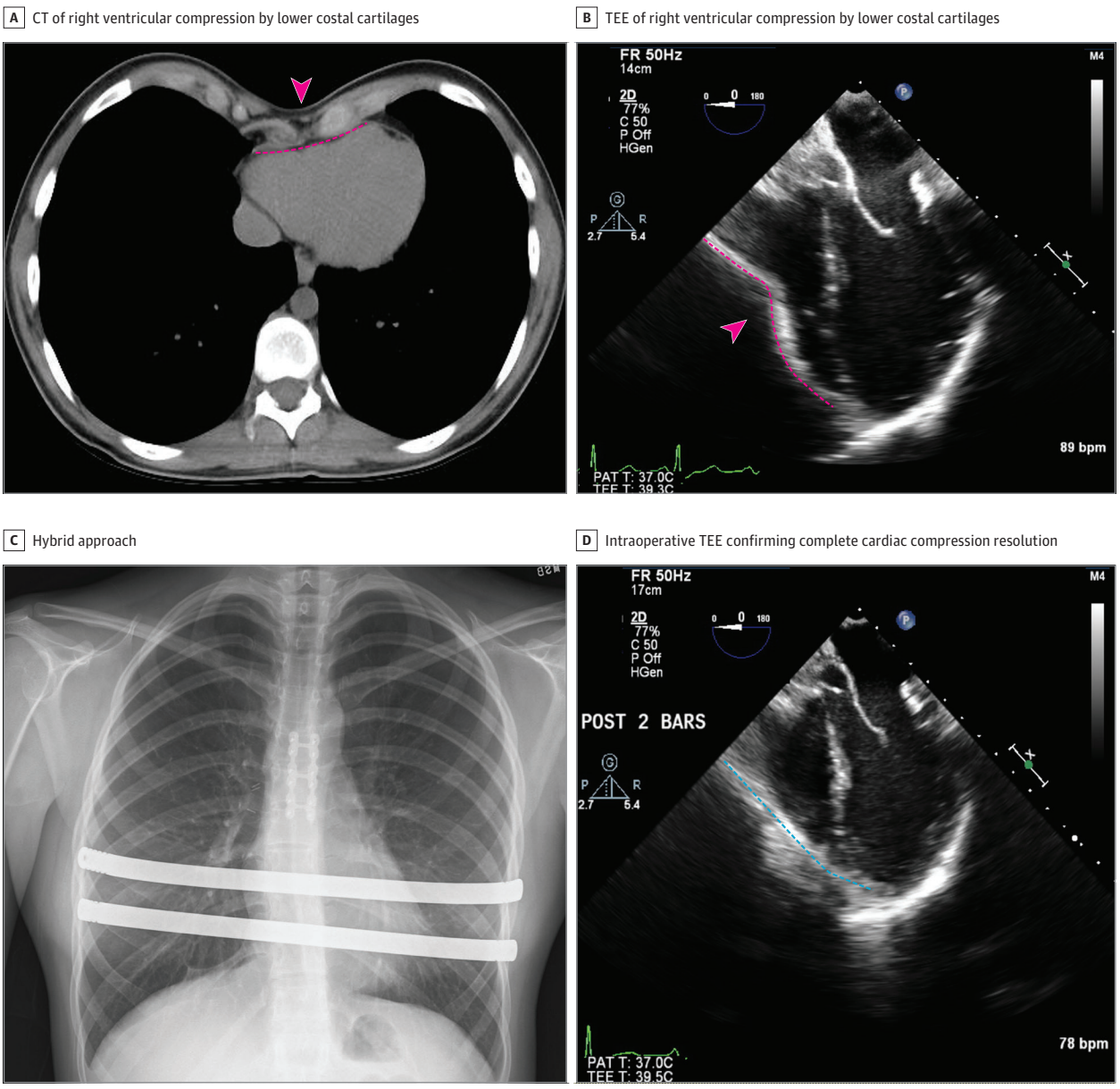
No special pain management strategies are needed beyond standard pectus protocols. Intercostal cryoablation[24] reduced epidural anesthesia use[64] and shorten hospital stays.[12,17]

Discussion

The one consistency in arcuatum literature is the variability in nomenclature and classification, leading to inconsistency in identification and treatment recommendations. Recognizing arcuatum as a separate entity is essential due to its origin, features, and unique management, a concept acknowledged by some.[11,12,18,71] Misclassification has been extensive: Ravitch misidentifying arcuatum,[38] Lester's initial diagnosis of arcuatum was excavatum,[14] later corrected,[2] but still considered a carinatum variant,[14] and more recent papers classifying arcuatum as an excavatum variant[1,6,8,16,40,57] or a mixed deformity.[8,72]

Arcuatum and superior carinatum are used interchangeably, highlighting confusion in the literature. Superior pectus carinatum features high sternal protrusion but lacks arcuatum's hallmark: fused, short, broad sternum supporting its sternal origin. Although once grouped with type II carinatum and considered a cartilaginous anomaly, Torre et al[73] modified Acastello's classification, distinguishing type II carinatum (cartilaginous) from arcuatum (sternal). Unlike true type II carinatum, arcuatum is often associated with lower sternal depression and frequently overlooked.[74] Unlike excavatum,

Figure 4. Radiographic Studies and Intraoperative Transesophageal Echocardiogram (TEE) Demonstrating Cardiac Compression Before Surgery and Its Complete Resolution After Hybrid Repair



Computed tomography (CT) (A) and TEE (B) showing right ventricular compression (pink arrowhead and dotted line) by the lower costal cartilages. This resolved after the hybrid approach (C), as demonstrated by intraoperative TEE (D) (blue dotted line).

arcuatum typically has normal Haller Index despite some depression, which can cause cardiac compression and symptoms.[3,12,14,24] Arcuatum is not merely a coexistence of excavatum and carinatum but a unique deformity with varying protrusions and depressions.[11,12,18,71]

History taking and a workup, including cardiopulmonary testing in symptomatic patients, is recommended. Arcuatum's prominent upper sternum is harder to conceal than excavatum, contributing to its significant psychosocial impact and possibly explaining the near-equal sex distribution, as breast tissue masking is less effective. Labeling arcuatum as purely cosmetic overlooks its physiologic potential.[16,61] The incidence of symptomatic patients remains unclear for multiple reasons. The deformity is likely underdiagnosed, and even when recognized, many believe it to be purely cosmetic due to its relatively normal Haller Index,[17,59,64] a factor affecting physicians' clinical assessment and decisions for ordering testing. Symptoms can be nonspecific, with the most common being exercise-associated dyspnea, chest pain, and tachycardia. Experienced surgeons noted symptom resolution after surgical repair.[6,12,19]

Though early literature highlights cardiopulmonary effects,[6,38] strong evidence remains limited. A recent study by Zeineddine et al[12] underscored dynamic cardiac compression from crowded lower costal cartilages during positional changes (eg, leaning

forward), offering insight into physiologic symptoms. Thorough history-taking and follow-up after management can help link symptoms to their true etiology and better documents surgical outcomes.

An important association in literature is CHD in patients with abnormal sternal ossification, particularly arcuatum.[1,60,68,75,76] Although Currarino-Silverman first described this syndrome by presenting cases with CHD[56] and patterns exist (Table), including CHD as diagnostic criteria is inaccurate, as many confirmed cases lack this association.[24,51] Preoperative echocardiogram is recommended, as CT alone may miss functional compromise and does not screen for CHD.[16,59,64]

Connective tissue disease associations exist (Table) but remain poorly supported.[16] Studies often exclude such patients,[57] report them as mixed deformities without clarification,[77] or fail to establish such association. Still, targeted evaluation is recommended when history or physical findings suggest such conditions.[12,17]

Surgical management has traditionally included modified Ravitch techniques,[16] but the hybrid approach, combining minimally invasive repair of pectus excavatum (MIRPE) with a midline incision to correct sternal protrusions and abnormal costochondral insertions, is gaining traction.[12,17,24,72] Recent hybrid techniques emphasize using bars to correct lower depression and enable superior, less invasive, correction.[12,17,24] Main differences lie in surgical sequence. St-Louis et al[17] reported inserting dissectors first, to delineate incisions, whereas Zeineddine et al[12] began with the open portion to minimize resisting forces and optimize reapproximation. St-Louis et al[17] performed cartilage resections before sternal wedge osteotomies. Others start with sternal osteotomy and fixation before cartilage reapproximation to minimize excision.[12] Many surgeons recommend rigid sternal fixation to prevent malunion and minimize pain.[12,24,64,78] St-Louis et al[17] felt Nuss bars eliminated the need for sternal plating and avoided vertical incisions for better aesthetics.[78,79]

Unlike excavatum, arcuatum may not require multiple bars, even in adults.[12,17] Due to the shortened sternum, bars may need to be placed below the sternal end, especially if lower costal cartilages are compressing the heart. Combined fixation methods of these lower bars are necessary to minimize migration risks.[12,19]

Regardless of an open or minimally invasive approach, the critical step is the wedge sternal osteotomy.[12,17,33] The unique sternal angulation and rigidity render MIRPE alone insufficient and may exaggerate angulations.[40,71] Multilevel osteotomies are usually not necessary to complete repair; a single wedge often suffices, preserving sternal integrity.[12,17,33,58,64] Determining the optimal wedge angle and width for realignment can be challenging and patient specific. Starting with a minimal cut width and expanding as needed is recommended.[12] Others suggest autologous bone/cartilage grafts to fill residual gaps.[24,43,51] Recent computer-aided design modeling innovations offer patient-specific cutting templates and may reduce surgical time.[32,65,80]

Extensive subperiosteal cartilage resection is unnecessary. In adults, heavily calcified cartilages may preclude subperiosteal dissection, making minimal cartilage resection essential. Rigid rib-to-sternal fixation is recommended in some cases.[12,19]

Limitations

This study has some limitations. Despite an extensive search, some articles on arcuatum may have been missed. Stratification was difficult, as many mixed-cohort studies did not differentiate arcuatum-specific findings, preventing data inclusion. Single-reviewer data extraction may introduce bias despite adherence to a standardized protocol to minimize this risk. Among included studies, 56.9% were small cohort studies, 27.7% case reports, 12.3% case series, and 3.1% technical reports, limiting generalizability. About 6.3% contained low-accuracy data but were included to ensure comprehensive coverage given the limited, heterogeneous literature.

Conclusions

This systematic review reveals that pectus arcuatum is an underrecognized and often misdiagnosed pectus deformity that is an entity distinct from carinatum and excavatum. Unified terminology and standardized diagnostic criteria are vital to improve management strategies and document surgical outcomes. Minimally invasive hybrid approaches continue to refine outcomes, balancing invasiveness with efficacy.

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