



OPEN ACCESS

EDITED BY

Massimo Bonacchi,
University of Florence, Italy

REVIEWED BY

Hendrik Tevaearai Stahel,
University Hospital of Bern, Switzerland

*CORRESPONDENCE

Jose A. Figueroa-Sanchez
✉ dr.figueroa@tec.mx

[†]These authors have contributed equally to
this work and share first authorship

RECEIVED 10 January 2025

ACCEPTED 22 October 2025

PUBLISHED 17 November 2025

CITATION

Martinez HR, Salazar-Alejo M, Ballesteros-
Suarez A, Perez-Martinez LE, Moran-
Guerrero JA, Flores-Salcido RE, Jimenez-
Franco V, Britton-Robles SC, Benvenuti-
Regato M and Figueroa-Sanchez JA (2025)
Postcoarctectomy syndrome: a contemporary
systematic review.
Front. Surg. 12:1518720.
doi: 10.3389/fsurg.2025.1518720

COPYRIGHT

© 2025 Martinez, Salazar-Alejo, Ballesteros-
Suarez, Perez-Martinez, Moran-Guerrero,
Flores-Salcido, Jimenez-Franco, Britton-
Robles, Benvenuti-Regato and Figueroa-
Sanchez. This is an open-access article
distributed under the terms of the [Creative
Commons Attribution License \(CC BY\)](#). The
use, distribution or reproduction in other
forums is permitted, provided the original
author(s) and the copyright owner(s) are
credited and that the original publication in
this journal is cited, in accordance with
accepted academic practice. No use,
distribution or reproduction is permitted
which does not comply with these terms.

Postcoarctectomy syndrome: a contemporary systematic review

Hector R. Martinez^{1,2†}, Misael Salazar-Alejo^{1,2†},
Ana Ballesteros-Suarez¹, Luis E. Perez-Martinez^{1,2},
Jose A. Moran-Guerrero^{1,2}, Rogelio E. Flores-Salcido²,
Vicente Jimenez-Franco^{1,3}, Silvia Cecilia Britton-Robles^{1,3},
Mario Benvenuti-Regato^{1,2} and Jose A. Figueroa-Sanchez^{1,2*}

¹Tecnologico de Monterrey, Escuela de Medicina y Ciencias de la Salud, Monterrey, Mexico, ²Instituto de Neurología y Neurocirugía, Centro Médico Zambrano Hellion, TecSalud, San Pedro Garza García, Mexico, ³Instituto de Cardiología y Medicina Vascular, Centro Médico Zambrano Hellion, TecSalud, San Pedro Garza García, México

Postcoarctectomy syndrome is a serious but poorly understood complication that can occur following surgical or endovascular repair of coarctation of the aorta. This condition presents with severe abdominal pain, persistent hypertension, and gastrointestinal symptoms that can be life-threatening if not promptly recognized and treated. Despite being a known complication for decades, the underlying mechanisms of postcoarctectomy syndrome remain unclear, creating challenges for both early diagnosis and effective treatment. We present the case of a patient with postcoarctectomy syndrome and a systematic review analyzing the clinical presentation, diagnostic approach, and treatment strategies for this complication. We also conduct a systematic literature review, identify key knowledge gaps, and propose future research priorities to address them.

KEYWORDS

vascular medicine, vascular surgery, cardiothoracic surgery, adverse events, review—systematic

1 Introduction

Coarctation of the aorta (CoA) is a congenital heart disease characterized by a localized narrowing of the thoracic aorta. Most commonly, the coarctation occurs near the insertion of the ductus arteriosus, distal to the left subclavian artery. However, some patients present with more extensive narrowing affecting longer aortic segments or tortuous areas and frequently occurs alongside other congenital heart defects (1). Clinical signs and symptoms may include a systolic or continuous murmur, weak or absent femoral pulses, hypertension, and aortic dissection (2). Patients may also experience headaches, epistaxis, and develop brain aneurysms (3, 4).

The first successful intervention for CoA was independently reported in 1945 by Crafoord & Nylin (5) and Gross & Hufnagel (6). Since this breakthrough, treatment options have expanded to include minimally invasive approaches such as balloon angioplasty and stent placement (7). Despite successful and timely repair, patients remain at risk for several complications, including persistent hypertension, recoarctation, aortic dissection, and postcoarctectomy syndrome (PCS) (1, 2).

PCS is a serious complication characterized by abdominal pain and systemic hypertension following surgical or endovascular repair of CoA. While PCS typically

develops within the first 7 days post-intervention, delayed presentation can occur weeks to months later. Patients may experience additional symptoms including vomiting, ileus, abdominal distension, fever, leukocytosis, and melena (3–7). Given that PCS can be life-threatening, clinicians must recognize this complication early and understand effective prevention and management strategies.

This review presents a representative case that demonstrates the clinical presentation, diagnostic approach, and treatment strategies for PCS. Additionally, we conduct a systematic review of current literature to synthesize existing knowledge, identify persistent knowledge gaps, and outline future research pathways to address them.

2 Case presentation

A Hispanic male in his late 40's initially presented 6 years prior to this report with an aneurysmal subarachnoid hemorrhage. During the aneurysm coiling procedure, digital subtraction angiography incidentally revealed CoA. After successfully managing the aneurysmal rupture, CT angiography of the aortic arch demonstrated a coarctation measuring 3.0×4.4 mm in diameter. At that time, CoA treatment was initially deferred to focus on neurological recovery and, despite medical advice, it was subsequently postponed based on the patient's preference (8).

Six years later, the patient returned with intermittent claudication. Blood pressure (BP) measurements revealed significant upper-to-lower extremity gradients and reduced ankle-brachial indices bilaterally. Despite treatment with 50 mg atenolol, 5 mg amlodipine, 160 mg valsartan, and 12.5 mg hydrochlorothiazide daily, the patient's BP remained elevated, averaging $\sim 150/106$ mmHg. Endovascular aortic repair was decided.

Prior to the procedure, all antihypertensive medications except atenolol were discontinued. Invasive hemodynamic measurements demonstrated a 60-mmHg transcoarctation gradient. A covered stent (BeGraft, 10×40 mm) was successfully implanted at the coarctation site. Post-deployment imaging confirmed proper stent expansion, with no evidence of aortic dissection or rupture. Immediate postoperative BP improved to 102/60 mmHg, with no residual gradient across the stent. Following postoperative protocol, the patient was admitted to the ICU where he remained hemodynamically stable on continued atenolol therapy. Two days post-procedure, the patient reported new-onset, colicky abdominal pain, rated 6/10 VAS in severity, primarily affecting the lower quadrants and radiated to the lower extremities. Associated symptoms included hyporexia, abdominal bloating, and nausea. Physical examination revealed a soft, non-distended abdomen with tenderness only on deep palpation. The pain remitted after a single 1-gram IV dose of acetaminophen. After a 24-h stay, the patient was discharged home. However, four days later, he returned with recurrent abdominal and back pain. CT angiography

demonstrated a patent stent in the descending aorta with no evidence of stent fracture, stenosis, aneurysm formation, or vascular dissection.

One month after the aortic repair, follow-up angiography showed continued stent patency with no evidence of leaks or restenosis. At the last follow-up, three months post-procedure, the patient remained asymptomatic, and atenolol therapy was discontinued.

3 Literature review

We conducted a systematic literature review following PRISMA guidelines (9). We performed a comprehensive literature search across PubMed, Web of Science, and Scopus databases for articles published from database inception through July 1st, 2025. Detailed electronic search strategies for each database are provided in [Supplementary Table SI](#). Two reviewers independently screened all identified titles and abstracts for relevance, followed by full-text assessment of potentially eligible studies against predetermined inclusion criteria. Studies were included if they reported on the incidence, pathophysiology, prevention, or treatment of PCS. We excluded editorials, commentaries, studies not specifically addressing PCS, and publications not available in English or Spanish. Any discrepancies between reviewers were resolved through discussion. Reference lists of included studies were screened to identify additional relevant publications. The flow of identified potential sources through the selection process is depicted in [Figure 1](#). Two reviewers independently performed quality appraisal using the JBI Critical Appraisal Tools for clinical studies and the SANRA scale for narrative reviews, depending on each study's design (10–12). Quality assessment results are reported in [Supplementary Tables SII–SVI](#).

4 Epidemiology

The reported incidence of PCS varies considerably across studies. The last compilation of incidence data was published in 1965 (3). Given this significant time gap, we sought to provide an updated compilation of reported PCS incidence rates. We identified all available studies reporting either PCS incidence or sufficient data to calculate incidence rates. To avoid case duplication, we cross-referenced authors and institutional affiliations across studies. Our analysis expanded the evidence base from 6 studies in the 1965 review to 37 studies, including two focused on catheter-based interventions, a treatment modality absent from previous reviews (13–42). We also compiled comprehensive symptom and mortality data, which were not detailed in the last compilation. [Table 1](#) summarizes the reported incidence of PCS from these studies. Mortality rate, which ranges from 0% to 3.1% in the largest studies, is detailed in [Supplementary Table SVII](#) along with information on reported symptom frequency.

TABLE 1 Reported incidence of postcoarctectomy syndrome.

Report	Publication year	CoA cases operated	Patients surviving	Patients with pcs	Percentage with pcs	Sample demographics
Brom ^a	1965	548	520	16	3.1%	Age: 0–1y (3.2%), 1–10y (19.7%), 10–20y (35.7%), 20–30 (20.9%), 30–60 (20.2%).
Koller et al. ^a	1987	362	362	0	0%	Age: 0–2y (20.4%), 2–19y (39.8%), ≥20y (39.8%).
Ibarra-Perez & Lillehei ^a	1969	Not reported	331	34	10.3%	219 males (66.1%) and 112 females (33.9%).
Lerberg et al. ^a	1982	334	329	32	9.7%	66.6% males, 28% were <1y, and 2.3% were African-american.
Toro-Salazar et al. ^a	2002	274	254	28	11%	Mean age at time of operation was 10.3 ± 9.5 years (median 7.3, range 0 to 41.5). Forty-seven patients were 1 year old at the initial operation, and 31 patients were 20 years old.
Pennington et al. ^a	1979	164	164	4	2.4%	Age: < 1y (9; 5.4%), 1–5y (17; 10.3%), 6–10y (20; 12.1%), 11–18y (42; 25.6%), 19–29y (35; 21.3%), 30–40y (14; 8.5%), > 40y (9; 5.4%).
Clarkson et al. ^a	1983	160	160	9	5.6%	67% males, Age: range 1–54y; 1–9y (40, 25%), 10–19y (62, 39%), 20–29y (26, 16%), 30–39y (16, 10%), 40–19y (11, 7%), ≥ 50 (5, 3%).
Chang & Burrington ^a	1972	194	159	13	8.2%	57 infants, 137 children and adults.
Braimbridge & Yen ^a	1965	119	112	4	3.5%	Mean age 22 years (range 3–56 years).
Palatianos et al. ^a	1985	107	107	0	0%	Mean age 6.4 years (range 4 days – 27 years), 65 males, 42 females.
Stansel et al. ^a	1977	100	99	3	3.0%	Age: 6–12 months (5, 5%), 1–5y (9, 9%), 5–10y (50, 50%), 10–20y (28, 28%), ≥ 20y (8, 8%).
Tawes et al. ^a	1970	103	98	8	8.2%	No details provided
Glancy et al. ^a	1983	70	67	4	5.9%	Mean age 17 years (range 1–49 years), 62 males, 22 females.
Cheatham et al. ^a	1979	80	77	2	2.6%	Mean age 4.5 years (range 1 week to 19 years), 51 males, 29 females. Forty percent were less than 2 years of age, whereas 60% were 2 years of age and older.
Sealy et al. ^{a,b}	1990	71	71	10	14.1%	No details provided
Patel et al. ^a	1977	65	65	5	7.7%	Age range 1–14 years, 30 males, 35 females.
Ring & Lewis	1956	73	64	18	28.1%	Mean age 11 years
Perianayagam et al. ^a	1980	51	46	0	0%	Age range 1 month to 48 years, 35 males and 16 females.
Wittig & Mulder ^a	1980	55	46	1	2.2%	Median age 3.6 months (range 1 day to 12 months), 32 males, 23 females.
Cleland et al.	1956	40	37	5	13.5%	Age range 1–55 years, 27 males, 13 females.
Trummer & Mannix	1963	33	33	8	24.2%	Mean age 11.9 years (range 4–33 years), 23 males, 10 females.
Mays & Sergeant	1965	35	33	4	12.1%	Mean age 10.8 years (range 10 days to 18 years), 22 males and 13 females.
Molaei et al. ^{c, a}	2011	26	26	2	7.7%	Age range 4–19 years.
Fox et al.	1980	25	25	0	0.0%	Age range 1 month to 30 years.
Lindensmith et al. ^a	1971	43	25	3	12%	Age range 2 weeks to 12 months.
Verska et al. ^a	1969	22	22	6	27.3%	Mean age 17.9 years (range 6–46 years), 17 males, 5 females.
Acevedo-Bañuelos et al. ^a	2013	20	20	0	0.0%	Mean age 85 days (range 1–180 days), 13 males, 7 females.
Bergdahl et al. ^a	1980	20	20	0	0%	There were 12 males with a mean age of 43 years (range 35–62 years) and 8 females with a mean age of 44 years (range 37–52 years).
Bergdahl et al. ^a	1980	19	19	1	5.2%	There were 14 boys with a mean age of 10 years (range 5–15 years) and 5 girls also with a mean age of 10 years (range 6–15 years).
Hurt & Hanbury	1957	17	17	2	11.8%	No details provided.
Moreno et al. ^a	1980	17	17	0	0%	Mean age 8.7 ± 5.44 years.
Srouji & Trusler ^a	1965	16	16	6	37.5%	Average age 7.3 years (range 4–15 years).
Reid & Dallachy	1958	15	15	1	6.7%	No details provided.

(Continued)

TABLE 1 Continued

Report	Publication year	CoA cases operated	Patients surviving	Patients with pcs	Percentage with pcs	Sample demographics
Parsons & Astley ^a	1966	13	10	2	20%	Age range 6 weeks to 13 years, 8 males, 2 females.
Rocchini et al. ^a	1976	7	7	5	71.4%	Mean age 9.2 years (range 7–14 years), 6 males, 1 female.
Kan et al.	1983	7	7	0	0%	Age range 10 months to 17 years, 6 males, 1 female.
Tefera et al. ^{c, a}	2016	3	3	1	33.3%	Mean age 10 years, 2 males, 1 female.

aNewly added studies since the last incidence compilation reported.

bUpdated report comprising the patients of the studies published in 1967 by the same single author.

cReport with patients exclusively treated with catheter-based intervention.

5 Risk factors

PCS shows strong male predominance, with males comprising 63–92.1% of cases (3, 36, 43). Large series have reported that, among patients who develop PCS, paradoxical hypertension precedes the syndrome in 25%–100% of cases (3, 14, 16, 26, 36, 40, 41, 44). Age at intervention may be a significant risk factor, with teenagers and adults with facing higher risk than infants (4, 13, 45). This discrepancy may be attributed to age-related factors, including abnormal aortic compliance, altered vasoreactivity, dysfunctional baroreceptors, and up-regulation of the renin-angiotensin system (RAS) (46). Regarding treatment approach, limited data exist comparing PCS incidence between endovascular and surgical repair.

6 Pathophysiology

PCS is believed to be the clinical manifestation of necrotizing arteritis that specifically affects vessels located distal to the previously coarcted aortic segment (3, 47–50). However, the exact mechanisms that trigger the development of these vascular lesions remain poorly understood, and multiple competing theories continue to be debated in the literature.

6.1 The link with paradoxical hypertension

A central paradigm suggests that PCS shares a common underlying cause with PH, which almost invariably precedes the syndrome (3, 6, 47). PH is a life-threatening, unexpected hemodynamic response that occurs despite successful surgical relief of CoA (39). PH follows a characteristic biphasic pattern. The first phase occurs within 24 h after aortic repair and presents as a brief spike in BP. The second phase develops 48–72 h after coarctectomy and is characterized by severe diastolic hypertension that typically coincides with the onset of the characteristic abdominal pain observed in PCS (22, 39, 51).

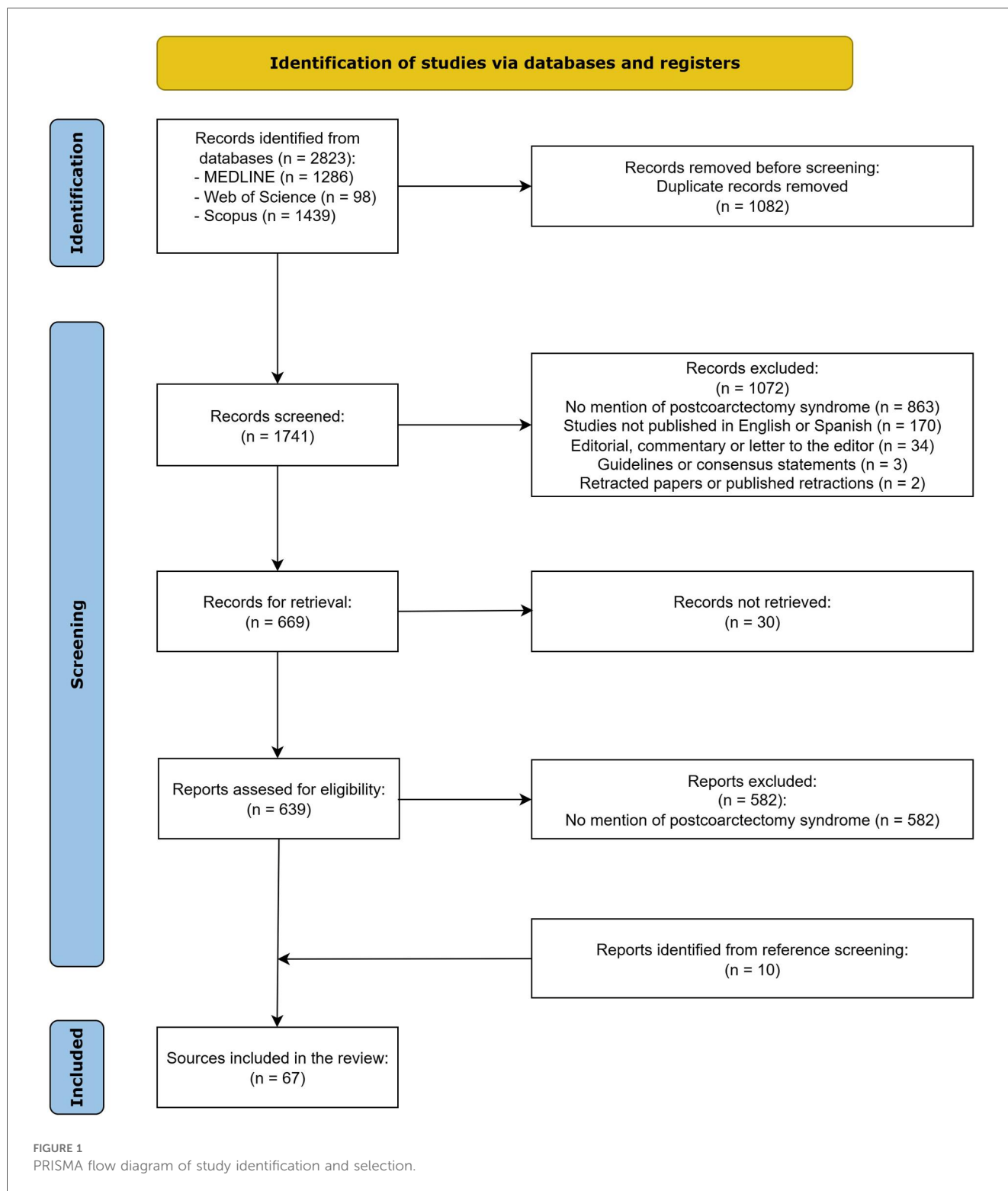
6.2 Sympathetic nervous system alterations

The elevation of catecholamines following coarctectomy suggests that sympathetic nervous system activation plays a

central role in the first phase of PH (42, 52, 53). The mechanism underlying this catecholamine surge appears to be directly linked to altered baroreceptor function. The primary arterial baroreceptors, located in the carotid sinus and aortic arch, are positioned proximal to the coarctation site. Consequently, both structures experience chronic exposure to long-lasting supranormal arterial pressures and gradually reset their regulatory setpoints to accommodate these abnormal hemodynamic conditions (54–56). Following successful coarctectomy, arterial pressure in the proximal aortic segment decreases abruptly. The chronically reset baroreceptors interpret this normal pressure reduction as pathologically low BP. In response, they decrease their inhibitory signals to the bulbar vasomotor centers, which then dramatically increase sympathetic output to compensate for what the altered baroreceptors perceive as hypotension. This compensatory response triggers excessive adrenaline and noradrenaline release (57–59). The elevation of catecholamines after surgery is not unique to coarctectomy, but the magnitude and duration of the noradrenaline surge are considerably greater compared to normotensive or hypertensive patients undergoing other surgical procedures (52, 57).

6.3 The role of the renin-angiotensin system

Another possible contributing mechanism involves the postsurgical increase in plasma renin activity and angiotensin II (AT-II). During the second phase of PH, there is a marked increase in both substances. This notable change in hormone concentrations is mediated by increased sympathetic nervous system activity following surgical aortic repair (60). The magnitude and duration of this hormonal response are uniquely characteristic of the postoperative period following coarctation repair (42). The severe abdominal pain and ileus characteristic of PCS may result from excessive mesenteric vasoconstriction induced by these substances, and the ensuing ischemia of the small bowel (3, 5, 38, 49). Given this pathophysiological understanding, RAS antagonists have been proposed as a promising therapeutic approach for controlling postoperative hypertension in this group of patients (61).



6.4 The mechanical hypothesis

An additional mechanism contributing to PCS involves the direct mechanical impact of surgical intervention and altered hemodynamics on downstream vasculature. Following aortic repair, the restoration of normal blood flow exposes mesenteric blood vessels to significantly increased pulsatile pressures,

potentially causing substantial arterial distension (57). Under these new hemodynamic conditions, the sudden exposure to higher pressures may result in excessive stimulation of viscerosensory fibers within vessel walls and subsequent arterial necrosis. This process culminates in the abdominal pain, distension, and ileus observed in PCS (4, 41, 47). Building on this mechanical theory, it has been suggested that the sudden

overdistension of small arteries and arterioles, previously adapted to lower-pressure blood flow, may trigger a protective vasospastic response. While this vasospasm could theoretically protect smaller vessels from further pressure-related damage, it may simultaneously induce bowel ischemia, ultimately causing the gastrointestinal manifestations characteristic of PCS (62, 63).

6.5 Experimental evidence for the mechanical hypothesis

The theory that relief of CoA exposes downstream vasculature to increased intra-arterial tension, thereby causing vascular damage, has substantial experimental support. This mechanical explanation for vascular lesions is derived from well-established animal models originally developed to study experimental and malignant hypertension (64–66). Among these experimental studies, Mays & Sergeant (3) highlight the model developed by Byrom & Dodson (66). In this landmark experiment, researchers forcibly injected Ringer's solution into rat carotid arteries, mechanically simulating the sudden increase in intra-arterial tension that occurs following relief of aortic obstruction (66). The resulting arterial lesions were remarkably similar to those described in numerous PCS case reports (8, 38, 55, 56). The experimental results support the concept that intravascular tension approaching the tolerance limits of exposed vasculature can cause overstretching of vessel walls and subsequent necrosis of the muscular layer. These weakened areas provide entry points for blood infiltration, which is then converted to fibrin through thromboplastin released from necrotic muscle cells. This initial damage triggers inflammatory cell infiltration of affected vessels as part of the natural repair process to restore vessel wall integrity (3, 66, 67).

6.6 Susceptibility of the mesenteric circulation

An intriguing observation is that, although multiple organs and vascular structures experience similar hemodynamic changes before and after coarctation repair, the splanchnic circulation, particularly the mesenteric vascular territories, demonstrates disproportionately severe involvement (3). The increased susceptibility of mesenteric vessels is theorized to result from two key factors. First, there is a relative lack of surrounding supportive tissue compared to other vascular beds. Second, these vessels possess inherent fragility due to chronic exposure to only subnormal pulse pressures prior to aortic repair (3, 6, 48, 68).

7 Prevention

Current strategies to prevent PCS include graduated serial stent dilation, early recognition and treatment of PH, and beta-blocker prophylaxis (45, 54, 69). However, emphasis on beta-blocker prophylaxis has decreased considerably, likely due to

significant improvements in postoperative antihypertensive therapy in the ICU since the 1970s (70). This progressive improvement was motivated by the need for alternatives to classic medications such as reserpine and trimethaphan, which were used to manage PH. One of the first major advances was the introduction of non-selective beta-blockers, particularly propranolol, for postoperative PH control (59, 71). Positive results with propranolol increased interest in adrenergic blockers for perioperative BP control in this patient population. Subsequently, esmolol, a selective β_1 -blocker, proved safe and effective for controlling postoperative hypertension in postcoarctectomy patients. Esmolol offered advantages over propranolol, including shorter duration of action and selective adrenoreceptor blockade (72, 73). However, beta-blockers carry a potential disadvantage. Although rare, unopposed beta-blockade with elevated catecholamines can stimulate alpha-adrenergic receptors, potentially causing the mesenteric ischemia characteristic of PCS (58). This limitation has led to recent trials exploring alternative approaches. A retrospective study (46) evaluated the effectiveness of intravenous labetalol, a combined selective α_1 - and non-selective β -antagonist, compared to no antihypertensive therapy in pediatric postcoarctectomy patients (46). The study demonstrated that intravenous labetalol was generally safe, effective, fast-acting, and easily convertible to oral therapy when continued treatment was necessary. These findings align with results reported in a small cohort study, which achieved adequate BP control using considerably smaller nitroprusside doses than previously required (58). Despite these advances, treatment approaches remain heterogeneous due to absent standardized guidelines and numerous available pharmacotherapy options (74).

8 Treatment

PCS typically resolves following a period of bowel rest and administration of antihypertensive therapy (3, 4, 67). Antihypertensive medications should be initiated immediately upon onset of PH and abdominal pain, using adequate dosages to control both conditions effectively (36). These medications serve dual therapeutic purposes: they reduce mechanical stress on mesenteric arterial walls caused by increased pulsatile blood flow and mitigate the vasoconstrictive and blood-shunting effects of the RAS (6). Additionally, some authors have reported positive outcomes using intravenous papaverine, a non-specific muscle relaxant, as an adjunctive treatment for PCS (5, 75). [Supplementary Table SVII](#) provides general information on PCS management as described in sources that documented specific treatments and patient outcomes. Among 121 patients for whom treatment details were reported, symptom duration ranged from 3 to 42 days. Thirteen patients required surgical intervention via laparotomy, and 4 deaths were reported (3, 5, 15, 16, 24, 36, 40, 41, 47, 62, 68, 75). Conservative management represented the most frequently employed therapeutic approach, reported in 75% of studies (3, 5, 15, 16, 41, 47, 62, 68, 75). Conservative measures typically included nil per os, intravenous fluid

administration, intubation, and gastric suction. Regarding antihypertensive therapy, reserpine was the most commonly used medication, reported in 30% of studies, followed by hydralazine, which was utilized in 15% of studies (3, 36, 39, 40, 68).

9 Discussion

Our understanding of PCS pathophysiology has evolved significantly since early hypotheses were proposed, particularly with advancements in splanchnic circulation physiology. The splanchnic vasculature, including its mesenteric circulation component, is regulated by both intrinsic and extrinsic control mechanisms (76–78). The intrinsic system operates through two primary pathways: metabolic and myogenic. The metabolic pathway induces vasodilation in response to tissue hypoxia and accumulation of metabolic byproducts. The myogenic pathway involves the Bayliss effect, a phenomenon of vasoconstriction that occurs when vascular wall myocytes undergo mechanical stretching. Conversely, decreased intravascular pressure triggers vasodilation, thereby maintaining relatively stable blood flow (78). The extrinsic system encompasses neurohumoral control, autonomic autoregulation, and central cardiovascular control. The neurohumoral component represents a balance between vasoconstrictors such as catecholamines and AT-II, and vasodilators including bradykinin, prostaglandins, and histamine (77, 78). An imbalance in these regulatory systems could explain the clinical findings observed in PCS. As previously discussed, altered baroreceptor setpoints, direct aortic arch manipulation, or both factors can considerably increase circulating catecholamines. These hormones activate β_1 -receptors on the juxtaglomerular apparatus and increase renin secretion, resulting in elevated AT-II levels (79). Both noradrenaline and AT-II produce exaggerated vasoconstriction in the mesenteric vasculature, potentially redistributing up to 80% of splanchnic blood flow and contributing substantially to ischemia in this vascular territory (80). Additionally, increased pulsatile pressure in vessels distal to the repaired coarctation could trigger exaggerated myogenic pathway activation, further reducing blood flow and promoting mesenteric ischemia. This abnormal response appears confined to the mesenteric circulation and may result from differences in adrenoceptor concentration and subtype ratios between organ systems. Specifically, predominant α -mediated vasoconstriction in the mesenteric vasculature could redistribute blood flow away from this territory (80).

Regarding identified knowledge gaps, further research into prevention and treatment of PH is warranted. Building on experiments by Price et al. (81) and findings by Siersma et al. (46) and Charlton et al. (58), prospective randomized studies could compare labetalol with different antihypertensive agents. Such studies should employ pre-specified therapy initiation thresholds, consider higher doses than previously reported, and include metrics such as adverse effect rates, PCS incidence, ICU length of stay, and treatment costs to refine the preventive and therapeutic roles of these medications (82). Similarly, using RAS-antagonists as alternative or adjunct pharmacological

management was proposed decades ago, but limited research has explored their therapeutic potential (83).

Notably, patients treated with endovascular techniques appear to have a less prominent sympathetic response, potentially decreasing PH risk (60). This observation warrants further investigation through prospective trials with larger sample sizes and inclusion of PCS incidence as an outcome measure. Additionally, several studies (68) have reported that patients may experience complications even months after coarctectomy (3, 68). Therefore, extended and close follow-up is warranted in future studies to promptly diagnose and address these delayed complications.

The considerable variability in PCS incidence rates across studies may result from differences in patient age, CoA treatment modalities, BP control protocols, and other risk factors that vary significantly between study populations. Future research should investigate how these factors influence PCS incidence rates.

The presented case underscores the need for continued clinical vigilance regarding PCS development. Even with optimal blood pressure management using β -blockers and endovascular techniques, this life-threatening complication can emerge days to weeks after CoA repair. Successful management requires rapid recognition and immediate attention to blood pressure control, pain management, and early assessment for potential surgical intervention. Long-term follow-up remains crucial, as patients may develop significant complications following PCS.

10 Conclusion

PCS remains an incompletely understood phenomenon with significant clinical implications. This article provides a comprehensive description of current pathophysiological understanding and an updated compilation of incidence data and treatment strategies reported in the literature. Clinical trials investigating novel and complementary pharmacological approaches could substantially improve our understanding of the underlying mechanisms driving PCS and provide a stronger evidence-based foundation for treatment decisions. Given that this complication can be fatal and carries considerable morbidity, healthcare providers treating patients with CoA must remain vigilant throughout the perioperative and extended follow-up periods. Active prevention strategies, prompt recognition and treatment of symptoms, and appropriate long-term follow-up are essential components of care for patients at risk of experiencing PCS. Early intervention and sustained clinical awareness can significantly impact patient outcomes and reduce the morbidity and mortality associated with this serious complication.

Author contributions

HM: Writing – original draft, Writing – review & editing, Conceptualization, Formal analysis, Investigation, Methodology, Project administration, Resources, Supervision, Validation. MS-A: Conceptualization, Data curation, Formal analysis,

Investigation, Methodology, Project administration, Resources, Software, Supervision, Validation, Visualization, Writing – original draft, Writing – review & editing. AB-S: Conceptualization, Data curation, Investigation, Methodology, Project administration, Software, Validation, Visualization, Writing – original draft, Writing – review & editing. LP-M: Conceptualization, Data curation, Investigation, Methodology, Project administration, Resources, Software, Validation, Visualization, Writing – original draft, Writing – review & editing. JM-G: Conceptualization, Data curation, Formal analysis, Investigation, Methodology, Project administration, Software, Validation, Visualization, Writing – original draft, Writing – review & editing. RF-S: Conceptualization, Data curation, Investigation, Methodology, Project administration, Software, Validation, Visualization, Writing – original draft, Writing – review & editing. VJ-F: Supervision, Validation, Writing – original draft, Writing – review & editing. SB-R: Supervision, Validation, Writing – original draft, Writing – review & editing. MB-R: Supervision, Validation, Writing – original draft, Writing – review & editing. JF-S: Writing – original draft, Writing – review & editing, Conceptualization, Data curation, Formal analysis, Investigation, Methodology, Project administration, Resources, Software, Supervision, Validation.

Funding

The author(s) declare that no financial support was received for the research and/or publication of this article.

Conflict of interest

The authors declare that the research was conducted in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.

References

- Kim YY, Andrade L, Cook SC. Aortic coarctation. *Cardiol Clin.* (2020) 38:337–51. doi: 10.1016/j.ccl.2020.04.003
- Dijkema EJ, Leiner T, Grotenhuis HB. Diagnosis, imaging and clinical management of aortic coarctation. *Heart.* (2017) 103:1148–55. doi: 10.1136/heartjnl-2017-311173
- Mays E, Sergeant C. Postcoarctectomy syndrome. *Arch Surg.* (1965) 91:58. doi: 10.1001/archsurg.1965.01320130060008
- Stewart D, Shores D, Alaish SM. Organ system response to cardiac function—splanchnic. *Elsevier.* (2019):150–159.e4. doi: 10.1016/b978-1-4557-0760-7.00015-2
- Pomar JL, Pelletier C, Hudon G, Hebert Y. Mesenteric arteritis complicating surgical repair of coarctation of the aorta. *Chest.* (1982) 82:508–10. doi: 10.1378/chest.82.4.508
- Fox S, Pierce WS, Waldhausen JA. Pathogenesis of paradoxical hypertension after coarctation repair. *Ann Thorac Surg.* (1980) 29:135–41. doi: 10.1016/s0003-4975(10)61651-7
- Fisher A, Benedict CR. Adult coarctation of the aorta: anaesthesia and postoperative management. *Anaesthesia.* (1977) 32:533–8. doi: 10.1111/j.1365-2044.1977.tb10000.x
- Figuerola-Sanchez J, Ferrigno A, Martinez-Rodriguez H. Multiple aneurysms, aortic coarctation, and persistent trigeminal artery. A unique presentation. *Surg Neurol Int.* (2019) 10(6). doi: 10.4103/sni.sni_395_18
- Page MJ, McKenzie JE, Bossuyt PM, Boutron I, Hoffmann TC, Mulrow CD, et al. The PRISMA 2020 statement: an updated guideline for reporting systematic reviews. *Br Med J.* (2021) 372:n71. doi: 10.1136/bmj.n71
- Baethge C, Goldbeck-Wood S, Mertens S. SANRA—a scale for the quality assessment of narrative review articles. *Res Integr Peer Rev.* (2019) 4(5). doi: 10.1186/s41073-019-0064-8
- Moola S, Munn Z, Tufanaru C, Aromataris E, Sears K, Sftcu R, et al. Chapter 7: systematic reviews of etiology and risk. In: Aromataris E, Munn Z, editors. *JBI Manual for Evidence Synthesis*. Adelaide: JBI (2020). <https://synthesismanual.jbi.global>
- Munn Z, Barker TH, Moola S, Tufanaru C, Stern C, McArthur A, et al. Methodological quality of case series studies: an introduction to the JBI critical appraisal tool. *JBI Database System Rev Implement Rep.* (2019) 18:2127–33. doi: 10.11124/jbisrir-d-19-00099
- Koller M, Rothlin M, Senning A. Coarctation of the aorta: review of 362 operated patients. Long-term follow-up and assessment of prognostic variables. *Eur Heart J.* (1987) 8:670–9. doi: 10.1093/eurheartj/8.7.670
- Lerberg DB, Hardesty RL, Siewers RD, Zuberbuhler JR, Bahnson HT. Coarctation of the aorta in infants and children: 25 years of experience. *Ann Thorac Surg.* (1982) 33:159–70. doi: 10.1016/s0003-4975(10)61903-0
- Tefera E, Leye M, Chanie Y, Raboisson M-J, Miro J. Percutaneous recanalization of totally occluded coarctation of the aorta in children using

Generative AI statement

The author(s) declare that Generative AI was used in the creation of this manuscript. Generative AI, specifically Claude Sonnet 4, was used to improve the grammatical structure, cohesion, and clarity of the text. Grammarly was also utilized to enhance language precision. No AI tools were used in the generation or interpretation of scientific content, data analysis, or conclusions. The core scientific findings, methodology, and intellectual contributions are entirely the work of the authors.

Any alternative text (alt text) provided alongside figures in this article has been generated by Frontiers with the support of artificial intelligence and reasonable efforts have been made to ensure accuracy, including review by the authors wherever possible. If you identify any issues, please contact us.

Publisher's note

All claims expressed in this article are solely those of the authors and do not necessarily represent those of their affiliated organizations, or those of the publisher, the editors and the reviewers. Any product that may be evaluated in this article, or claim that may be made by its manufacturer, is not guaranteed or endorsed by the publisher.

Supplementary material

The Supplementary Material for this article can be found online at: <https://www.frontiersin.org/articles/10.3389/fsurg.2025.1518720/full#supplementary-material>

brockenbrough needle and covered stents. *Ann Pediatr Cardiol.* (2016) 9:153. doi: 10.4103/0974-2069.180664

16. Stansel HC, Tabry IF, Poirier RA, Berman MA, Hellenbrand WE. One hundred consecutive coarctation resections followed from one to thirteen years. *J Pediatr Surg.* (1977) 12:279–86. doi: 10.1016/0022-3468(77)90002-1

17. Acevedo-Bañuelos I, González-Peña J, Chagolla-Santillán MÁ, Hernández-Morales G, Fariás-Serratos CV. Reparación quirúrgica de coartación aórtica en lactantes menores. *Archivos de Cardiología de México.* (2013) 83:159–64. doi: 10.1016/j.acmx.2013.03.004

18. Molaei A, Merajie M, Mortezaeian H, Malakan Rad E, Haji Heidar Shemirani R. Complications of aortic stenting in patients below 20 years old: immediate and intermediate follow-up. *J Tehran Heart Cent.* (2011) 6:202–5.

19. Toro-Salazar OH, Steinberger J, Thomas W, Rocchini AP, Carpenter B, Moller JH. Long-term follow-up of patients after coarctation of the aorta repair. *Am J Cardiol.* (2002) 89:541–7. doi: 10.1016/s0002-9149(01)02293-7

20. Palatianos GM, Kaiser GA, Thurer RJ, Garcia O. Changing trends in the surgical treatment of coarctation of the aorta. *Ann Thorac Surg.* (1985) 40:41–5. doi: 10.1016/s0003-4975(10)61167-8

21. Clarkson PM, Nicholson MR, Barratt-Boyes BG, Neutze JM, Whitlock RM. Results after repair of coarctation of the aorta beyond infancy: a 10 to 28 year follow-up with particular reference to late systemic hypertension. *Am J Cardiol.* (1983) 51:1481–8. doi: 10.1016/0002-9149(83)90661-6

22. Glancy DL, Morrow AG, Simon AL, Roberts WC. Juxtaductal aortic coarctation. *Am J Cardiol.* (1983) 51:537–51. doi: 10.1016/s0002-9149(83)80094-0

23. Kan JS, White RI, Mitchell SE, Farmlett EJ, Donahoo JS, Gardner TJ. Treatment of restenosis of coarctation by percutaneous transluminal angioplasty. *Circulation.* (1983) 68:1087–94. doi: 10.1161/01.cir.68.5.1087

24. Bergdahl L, Jonasson R, Björk VO. Late results of operation in children with coarctation of the aorta. *Scand J Thorac Cardiovasc Surg.* (1980) 14:83–9. doi: 10.3109/14017438009109859

25. Bergdahl L, Jonasson R, Björk VO. Late results of operation for coarctation of the aorta in patients more than 35 years of age. *Scand J Thorac Cardiovasc Surg.* (1980) 14:77–82. doi: 10.3109/14017438009109858

26. Pennington GD, Liberthson RR, Jacobs M, Scully H, Goldblatt A, Daggett WM. Critical review of experience with surgical repair of coarctation of the aorta. *J Thorac Cardiovasc Surg.* (1979) 77:217–29. doi: 10.1016/s0022-5223(19)40960-4

27. Moreno NN, Campo TD, Kaiser GA, Pallares VS. Technical and pharmacologic management of distal hypotension during repair of coarctation of the aorta. *J Thorac Cardiovasc Surg.* (1980) 80:182–6. doi: 10.1016/s0022-5223(19)37787-6

28. Perianayagam WJ, Muralidharan S, Jairaj PS, Krishnaswamy S, Sukumar IP, Cherian G, et al. Experience with the surgical treatment of coarctation of the aorta in India. *Thorax.* (1980) 35:950–2. doi: 10.1136/thx.35.12.950

29. Wittig JH, Mulder DG. Repair of coarctation of the aorta in infants. *Am J Surg.* (1980) 140:158–63. doi: 10.1016/0002-9610(80)90434-1

30. Cheatham JE, Williams GR, Thompson WM, Luckstead EF, Razook JD, Elkins RC. Coarctation: a review of 80 children and adolescents. *Am J Surg.* (1979) 138:889–93. doi: 10.1016/0002-9610(79)90317-9

31. Patel R, Singh SP, Abrams L, Roberts KD. Coarctation of aorta with special reference to infants. Long-term results of operation in 126 cases. *Heart.* (1977) 39:1246–53. doi: 10.1136/hrt.39.11.1246

32. Chang JHT, Burrington JD. Coarctation of the aorta in infants and children. *J Pediatr Surg.* (1972) 7:127–35. doi: 10.1016/0022-3468(72)90486-1

33. Lindesmith GG, Stanton RE, Stiles QR, Meyer BW, Jones JC. Coarctation of the thoracic aorta. *Ann Thorac Surg.* (1971) 11:482–97. doi: 10.1016/s0003-4975(10)65487-2

34. Parsons CG, Astley R. Recurrence of aortic coarctation after operation in childhood. *Br Med J.* (1966) 1:573–7. doi: 10.1136/bmj.1.5487.573

35. Brom GA. Narrowing of the aortic isthmus and enlargement of the mind. *J Thorac Cardiovasc Surg.* (1965) 50:166–80. doi: 10.1016/s0022-5223(19)33204-0

36. Ibarra-Perez C, Walton Lillehei C. Treatment of mesenteric arteritis following resection of coarctation of the aorta. *J Thorac Cardiovasc Surg.* (1969) 58:135–9. doi: 10.1016/s0022-5223(19)42641-x

37. Braimbridge MV. Coarctation in the Elderly. (1965).

38. Tawes RL, Bull JC, Roe BB. Hypertension and abdominal pain after resection of aortic coarctation. *Ann Surg.* (1970) 171:409–12. doi: 10.1097/0000658-197003000-00014

39. Sealy WC. Paradoxical hypertension after repair of coarctation of the aorta: a review of its causes. *Ann Thorac Surg.* (1990) 50:323–9. doi: 10.1016/0003-4975(90)90768-2

40. Verska JJ, De Quattro V, Woolley MM. Coarctation of the aorta. *J Thorac Cardiovasc Surg.* (1969) 58:746–53. doi: 10.1016/s0022-5223(19)42555-5

41. Srouji MN, Trusler CA. Paradoxical Hypertension and the Abdominal Pain Syndrome Following Resection of Coarctation of the Aorta. (1965) 92).

42. Rocchini AP, Rosenthal A, Barger AC, Castaneda AR, Nadas AS. Pathogenesis of paradoxical hypertension after coarctation resection. *Circulation.* (1976) 54:382–7. doi: 10.1161/01.cir.54.3.382

43. Cheitlin MD. Coarctation of the aorta. *Med Clin N Am.* (1977) 61:655–73. doi: 10.1016/s0025-7125(16)31323-2

44. Stemmer EA, Connolly JE. Mesenteric vascular insufficiency. Identification and management. *Calif Med.* (1973) 118:18–29.

45. Sapin SO, Rosengart RM, Salem MM. Chest pain during stenting of a native aortic coarctation: a case for acute intercostal muscle ischemia and rhabdomyolysis. *Catheter Cardiovasc Interv.* (2002) 57:217–20. doi: 10.1002/ccd.10293

46. Siersma C, Brouwer CNM, Sojak V, Ten Harkel ADJ, Roeleveld PP. Treatment of post-coarctectomy hypertension with labetalol—a 9-year single-center experience. *World J Pediatr Congenit Heart Surg.* (2022) 13:701–6. doi: 10.1177/2150135122111797

47. Ring DM, Lewis FJ. Abdominal pain following surgical correction of coarctation of the aorta: a syndrome. *J Thorac Surg.* (1956) 31:718–24. doi: 10.1016/s0096-5588(20)30881-3

48. Downing DF. Coarctation of the Aorta. *AMA J Dis Children.* (1958) 96(711):701–6. doi: 10.1001/archpedi.1958.02060060713010

49. Sealy WC. Coarctation of the aorta and hypertension. *Ann Thorac Surg.* (1967) 3:15–28. doi: 10.1016/s0003-4975(10)66683-0

50. Hurt RL, Hanbury WJ. Intestinal vascular lesions simulating polyarteritis nodosa after resection of coarctation of the aorta. *Thorax.* (1957) 12:258–63. doi: 10.1136/thx.12.3.258

51. Rothlin ME. Surgical repair of coarctation: early and late results. *J Intervent Cardiology.* (1992) 5:203–8. doi: 10.1111/j.1540-8183.1992.tb00428.x

52. Benedict CR, Grahame-Smith DG, Fisher A. Changes in plasma catecholamines and dopamine beta-hydroxylase after corrective surgery for coarctation of the aorta. *Circulation.* (1978) 57:598–602. doi: 10.1161/01.cir.57.3.598

53. Vyden JK. Subnormal amylase and gamma globulin levels in the postcoarctation abdominal pain syndrome. *J Thorac Cardiovasc Surg.* (1972) 63:439–41. doi: 10.1016/s0022-5223(19)41908-9

54. Matsuyama K, Sonoda E, Nakao K, Horio Y, Yasue H. Baroreceptor reflex in a patient with coarctation of the aorta. *Clin Cardiol.* (1987) 10:535–6. doi: 10.1002/clc.4960100915

55. Iglar FO, Boerboom LE, Werner PH, Donegan JH, Zuperku EJ, Bonchek LI, et al. Coarctation of the aorta and baroreceptor resetting. A study of carotid baroreceptor stimulus-response characteristics before and after surgical repair in the dog. *Circ Res.* (1981) 48:365–71. doi: 10.1161/01.res.48.3.365

56. Hanson E, Eriksson BO, Sivertsson R. Baroreceptor reflexes after coarctectomy. *Clinical Physiology.* (1981) 1:503–9. doi: 10.1111/j.1475-097x.1981.tb00917.x

57. Goodall MCC, Sealy WC. Increased sympathetic nerve activity following resection of coarctation of the thoracic aorta. *Circulation.* (1969) 39:345–51. doi: 10.1161/01.cir.39.3.345

58. Charlton GA, Ladd DR, Friesen RM, Friesen RH. Labetalol infusion attenuates paradoxical hypertension and decreases plasma renin activity after repair of coarctation of the aorta in children. *J Cardiothorac Vasc Anesth.* (2020) 34:3348–51. doi: 10.1053/j.jvca.2020.05.027

59. Gidding SS, Rocchini AP, Beekman R, Szpunar CA, Moorehead C, Behrendt D, et al. Therapeutic effect of propranolol on paradoxical hypertension after repair of coarctation of the aorta. *N Engl J Med.* (1985) 312:1224–8. doi: 10.1056/nejm198505093121904

60. Choy M, Rocchini AP, Beekman RH, Rosenthal A, Dick M, Crowley D, et al. Paradoxical hypertension after repair of coarctation of the aorta in children: balloon angioplasty versus surgical repair. *Circulation.* (1987) 75:1186–91. doi: 10.1161/01.cir.75.6.1186

61. Wilkinson C, Clark H. Refractory hypertension during coarctectomy. *Anesthesiology.* (1982) 57:540–2. doi: 10.1097/0000542-198212000-00020

62. Reid HC, Dallachy R. Infarction of ileum following resection of coarctation of the aorta. *Br J Surg.* (1958) 45:625–32. doi: 10.1002/bjs.18004519413

63. Moore JW, Lovett EJ, Kirby WC. Gastrointestinal hemorrhage after combined percutaneous angioplasty of aortic coarctation and valvuloplasty of aortic stenosis in an infant. *Pediatr Cardiol.* (1993) 14:53–5. doi: 10.1007/bf00794848

64. Fleming HA. Factors involved in the production of acute arterial lesions in rabbits with experimental renal hypertension. *J Pathol Bacteriol.* (1953) 65:441–50. doi: 10.1002/path.1700650216

65. Wilson C, Byrom FB. Renal changes in malignant hypertension. *Lancet.* (1939) 233:136–9. doi: 10.1016/s0140-6736(00)60143-5

66. Byrom FB, Dodson LF. The causation of acute arterial necrosis in hypertensive disease. *J Pathol Bacteriol.* (1948) 60:357–68. doi: 10.1002/path.1700600302

67. Hallman GL, Bloodwell RD, Cooley DA. Coarctation of the thoracic Aorta. *Surg Clin North America.* (1966) 46:893–9. doi: 10.1016/s0039-6109(16)37933-6

68. Trummer MJ, Mannix EP. Abdominal pain and necrotizing mesenteric arteritis following resection of coarctation of the aorta. *J Thorac Cardiovasc Surg.* (1963) 45:198–209. doi: 10.1016/s0022-5223(19)32884-3
69. Shirai T, Amano J, Fujii N, Hirabayashi S, Suzuki A. Rupture of Inferior phrenic artery aneurysm. *Chest.* (1994) 106:1290–1. doi: 10.1378/chest.106.4.1290
70. Love BA, Fischer GW, Stelzer P, Fuster V. Aortic coarctation in the adult. *Springer London.* (2013) 45:2521–49. doi: 10.1007/978-1-4471-4619-3_184
71. Will RJ, Walker OM, Traugott RC, Treasure RL. Sodium nitroprusside and propranolol therapy for management of postcoarctectomy hypertension. *J Thorac Cardiovasc Surg.* (1978) 75:722–4. doi: 10.1016/s0022-5223(19)41320-2
72. Tabbutt S, Nicolson SC, Adamson PC, Zhang X, Hoffman ML, Wells W, et al. The safety, efficacy, and pharmacokinetics of esmolol for blood pressure control immediately after repair of coarctation of the aorta in infants and children: a multicenter, double-blind, randomized trial. *J Thorac Cardiovasc Surg.* (2008) 136:321–8. doi: 10.1016/j.jtcvs.2007.09.086
73. Wiest DB, Garner SS, Uber WE, Sade RM. Esmolol for the management of pediatric hypertension after cardiac operations. *J Thorac Cardiovasc Surg.* (1998) 115:890–7. doi: 10.1016/s0022-5223(98)70371-x
74. Moffett BS, Penny DJ. Variability in treatment of post-coarctectomy hypertension: a multicenter study. *Pediatr Cardiol.* (2016) 37:772–7. doi: 10.1007/s00246-016-1349-z
75. Jintao F, Xingang W, Wenhui W, Wei M. Abdominal pain after stenting for aortic coarctation. *Journal of vascular surgery cases. Innov Techniq.* (2020) 6:678–80. doi: 10.1016/j.jvscit.2019.11.004
76. Harper D, Chandler B. Splanchnic circulation. *BJA Educ.* (2016) 16:66–71. doi: 10.1093/bjaceaccp/mkv017
77. Zimmerman P, Huseynova K, Pillai L. Anatomy and physiology of the mesenteric circulation. *Elsevier.* (2019) 58:1014–26. doi: 10.1016/b978-0-323-40232-3.00086-8
78. Hashmi S, Khattab A, Ehrenpreis ED. Physiology of the mesenteric circulation. *Springer International Publishing.* (2021) 6:107–19. doi: 10.1007/978-3-030-71963-0_13
79. Thomas GD. Neural control of the circulation. *Adv Physiol Educ.* (2011) 35:28–32. doi: 10.1152/advan.00114.2010
80. Fink GD, Osborn JW. The splanchnic circulation. *Elsevier.* (2012):211–3. doi: 10.1016/b978-0-12-386525-0.00043-3
81. Price HL, Cooperman LH, Warden JC. Control of the splanchnic circulation in man. *Circ Res.* (1967) 21:333–40. doi: 10.1161/01.res.21.3.333
82. Roeleveld PP, Zwijsen EG. Treatment strategies for paradoxical hypertension following surgical correction of coarctation of the aorta in children. *World J Pediatr Congenit Heart Surg.* (2017) 8:321–31. doi: 10.1177/2150135117690104
83. Casta A, Conti VR, Talabi A, Brouhard BH. Effective use of captopril in postoperative paradoxical hypertension of coarctation of the aorta. *Clin Cardiol.* (1982) 5:551–3. doi: 10.1002/clc.4960051009