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Congenital Pouch Colon: A Comprehensive Systematic Review and Meta-Analysis of Current Evidence A PRISMA-compliant synthesis of anatomical patterns, management strategies, and clinical outcomes

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Abstract

Background.

Congenital pouch colon (CPC) is a rare and severe variant of anorectal malformation in which part or all of the colon is replaced by a dilated pouch communicating with the urogenital tract via a fistula. Although reported worldwide, CPC shows striking geographical clustering in northern and north-western India. The literature is dominated by heterogeneous single-institution case series, and pooled estimates of its anatomical spectrum, management and outcomes are lacking.

Objectives.

To systematically synthesise the global evidence on CPC and to derive pooled estimates of subtype distribution, associated anomalies, perioperative mortality and long-term continence, and to appraise the evolution of surgical strategy.

Methods.

We searched PubMed, Scopus, Embase and Web of Science from inception to 31 December 2025 following PRISMA 2020. Observational studies (case series ≥ 5 patients and comparative studies) reporting extractable CPC data were eligible. Two reviewers independently screened records, extracted data and appraised methodological quality using the JBI checklist for case series. Proportions were pooled using random-effects (DerSimonian–Laird) models with logit transformation; heterogeneity was quantified with the I^2 statistic.

Results.

Forty-seven studies met inclusion criteria (34 contributing to meta-analysis; N = 1 926 patients). The pooled male-to-female ratio was 2.5:1. Type III (30.4%) and Type II (27.9%) were the commonest subtypes. Associated anomalies affected roughly half of patients, predominantly genitourinary (34.6%). The pooled perioperative mortality was 20.4% (95% CI 15.2–26.9; I^2 = 53%), declining from 41.0% in series published on or before 1990 to 8.9% after 2020. Good long-term faecal continence was achieved in 52% of assessed survivors overall, but only 38% in high (Type I–II) pouches versus 64% in low (Type III–IV) pouches.

Conclusions.

CPC carries substantial but steadily improving mortality and type-dependent functional outcomes. Coloplasty-based colon-preserving strategies and staged reconstruction have accompanied better

survival. Standardised classification, prospective multicentre registries and consensus outcome measures are urgently needed. This review is limited by the observational, single-region nature of the evidence base.

PROSPERO Registration:

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1. Introduction

Anorectal malformations (ARM) constitute one of the most frequent groups of congenital surgical anomalies of the neonate, occurring in approximately 1 in 4 000–5 000 live births[1]. Within this heterogeneous spectrum, congenital pouch colon (CPC) occupies a distinctive and clinically formidable position. In CPC, a variable length of the colon—and in the most severe forms the entire colon—is replaced by a dilated, poorly haustrated pouch that terminates in a fistulous communication with the lower urinary tract in males or the genital tract in females. The condition was first delineated as a discrete entity by Singh and Pathak in 1972[2], who described a series of neonates with a markedly foreshortened colon associated with imperforate anus, a constellation that later became widely known as “pouch colon syndrome”[3,4].

The epidemiology of CPC is remarkable for pronounced geographical clustering. The overwhelming majority of reported cases originate from northern and north-western India—particularly the states of Uttar Pradesh, Rajasthan, Bihar and the Delhi region—where CPC may account for 2–18% of all anorectal malformations in tertiary referral series[5,6,7], an order of magnitude higher than the sporadic reports emerging from other parts of the world. The reasons for this clustering remain unresolved and are variously attributed to environmental, nutritional, consanguineous and as-yet-undefined genetic factors, rendering CPC both a surgical challenge and an unresolved question in developmental biology.

Clinically, CPC presents in the neonatal period with features of low intestinal obstruction, absent anal opening and, frequently, the passage of meconium or gas in the urine reflecting the colovesical or colourethral fistula[6]. A characteristic radiographic sign—a single, very large gas-filled or air-fluid-containing viscus occupying much of the abdomen on the erect film[8,9]—should prompt immediate consideration of the diagnosis. The abnormal, ectatic pouch carries a precarious, often single-vessel blood supply and a poorly developed muscular wall[10], which together underlie both the operative difficulty and the historically high mortality of the condition.

Surgical management has evolved substantially over five decades. Early strategies centred on excision of the entire pouch with ileostomy or high colostomy[3,5], sacrificing colonic absorptive length and predisposing survivors to fluid and electrolyte derangement. Subsequent innovations—most notably colon-preserving tubular coloplasty and staged reconstruction[11,12] culminating in an abdomino-perineal

pull-through[13]—sought to balance safe removal of the abnormal pouch against preservation of functional bowel length and continence. Despite these advances, outcomes remain type-dependent and the evidence base is fragmented across numerous single-centre retrospective series that employ divergent classification schemes and outcome definitions.

To date, no quantitative synthesis has integrated this dispersed literature. Narrative reviews have summarised individual centres' experience[6,7,14], but pooled estimates of subtype frequency, the burden of associated anomalies, perioperative mortality and long-term functional outcome are unavailable, hindering evidence-based counselling and benchmarking. We therefore undertook a PRISMA-compliant systematic review and meta-analysis with the following objectives:

- To characterise the demographic and geographical distribution of reported CPC;
- To derive pooled estimates of the anatomical subtype distribution (Narasimharao classification) and the prevalence of associated congenital anomalies;
- To quantify pooled perioperative mortality and to examine its temporal trend;
- To summarise long-term faecal continence stratified by pouch severity; and
- To map the evolution of surgical strategy and identify priorities for future research.

2. Methods

2.1 Protocol, registration and reporting

This review was conducted and reported in accordance with the Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA 2020) statement[15] and the Meta-analysis Of Observational Studies in Epidemiology (MOOSE) guidance[16]. The protocol was registered prospectively in the PROSPERO international registry (CRD420261462539). No important deviations from the protocol occurred; any post-hoc analyses are identified as such.

2.2 Eligibility criteria

Studies were selected according to the following participant, exposure, comparator, outcome and study-design (PECOS) framework:

- **Population**: neonates, infants and children with a confirmed diagnosis of congenital pouch colon of any anatomical subtype.
- **Exposure/index condition**: CPC diagnosed operatively, radiologically or histopathologically.
- **Comparators**: where reported, alternative surgical strategies or CPC subtypes; comparators were not required for inclusion.
- **Outcomes**: subtype distribution, associated anomalies, operative approach, perioperative mortality, and long-term continence or functional outcome.

- **Study designs**: observational case series (≥ 5 patients), cohort and comparative studies. Single case reports, editorials, narrative reviews, conference abstracts without extractable data, and animal or cadaveric studies were excluded.

Where multiple publications reported overlapping cohorts from the same institution and period, the most complete or most recent report was retained to avoid double-counting; earlier reports were used only for non-overlapping variables.

2.3 Information sources and search strategy

We searched PubMed/MEDLINE, Scopus, Embase and Web of Science Core Collection from database inception to 31 December 2025, without language restriction. The search combined controlled vocabulary and free-text terms for the index condition and its historical synonyms. A representative PubMed string was:

("congenital pouch colon" OR "pouch colon" OR "pouch colon syndrome" OR "congenital short colon" OR "short colon") AND ("anorectal malformation" OR "imperforate anus" OR "anorectal agenesis" OR colon* OR neonat* OR infant*)

Reference lists of all included articles and of prior narrative reviews were hand-searched to identify additional eligible studies. The grey literature was scanned through Google Scholar for the first 200 results.

2.4 Study selection and data extraction

Records were de-duplicated in a reference manager. Two reviewers independently screened titles and abstracts, then assessed full texts against the eligibility criteria; disagreements were resolved by discussion or a third reviewer. A piloted, standardised extraction form captured: bibliographic details; country and study period; design; sample size; sex distribution; subtype distribution (as originally classified and, where possible, mapped to the Narasimharao scheme); associated anomalies; fistula type; operative procedures and staging; perioperative mortality; length of follow-up; and functional/continence outcomes. Extraction was performed independently in duplicate and cross-checked.

2.5 Methodological quality assessment

The methodological quality of included case series was appraised with the Joanna Briggs Institute (JBI) Critical Appraisal Checklist for Case Series[17] (ten items addressing inclusion criteria, condition measurement, case-ascertainment validity, consecutive and complete inclusion, demographic and clinical reporting, outcome and follow-up completeness, and appropriateness of statistical presentation). Each item was rated "yes," "no," "unclear" or "not applicable." Studies fulfilling ≥ 8 items were rated low risk of bias, 5–7 moderate, and ≤ 4 high. Quality was used to inform sensitivity analyses rather than as an exclusion filter.

2.6 Data synthesis and statistical analysis

Because outcomes were proportions drawn from single-arm observational studies with anticipated heterogeneity, we pooled event proportions using random-effects models (DerSimonian–Laird estimator)[18] after logit transformation, back-transforming pooled estimates and 95% confidence intervals for presentation. Between-study heterogeneity was quantified with the I^2 statistic[19] (interpreted as low ≤ 25%, moderate 26–75%, high > 75%) and the τ^2 variance, with Cochran's Q test. Pre-specified subgroup and meta-regression analyses examined publication era, pouch severity (high, Types I–II vs low, Types III–IV) and geographical origin. Small-study effects and publication bias were assessed by funnel-plot inspection and Egger's regression[20] where ≥ 10 studies contributed. Leave-one-out sensitivity analyses evaluated the influence of individual studies. A two-sided $p < 0.05$ was considered significant. Analyses were conducted in R (metafor/meta packages)[21]; figures were rendered at publication resolution.

3. Results

3.1 Study selection

The database searches yielded 1 284 records, supplemented by 46 records identified through hand-searching and reference screening. After removal of duplicates, 812 records underwent title and abstract screening, of which 691 were excluded as clearly irrelevant. Of 121 full-text articles assessed, 74 were excluded—most commonly because they were single case reports (n = 29), lacked extractable aggregate data (n = 21), reported overlapping cohorts (n = 14) or were non-English without extractable numerical data (n = 10). Forty-seven studies met the inclusion criteria for qualitative synthesis, of which 34 (pooled N = 1 926 patients) contributed to the quantitative meta-analysis. The selection process is summarised in Fig. 1.

3.2 Characteristics of included studies

The included studies spanned from 1972 to 2025 and were overwhelmingly retrospective single-institution case series. The great majority originated from India, with sporadic contributions from other regions confirming the entity's global rarity outside the South-Asian cluster. Sample sizes ranged from small consecutive series to large tertiary-centre cohorts exceeding 70 patients[5,22,23,24]. Table 1 presents the characteristics of a representative subset of the largest and most frequently cited contributing studies.

Table 1
Characteristics of representative included studies.

Study (first author, year)	Country	Design	Study period	N	Class.*
Singh & Pathak, 1972	India	Case series (retro.)	1966–71	16	–
Narasimharao et al., 1984	India	Case series (retro.)	1974–83	17	N
Budhiraja et al., 1996	India	Case series (retro.)	1988–94	27	N
Wakhlu et al., 1996	India	Case series (retro.)	1984–94	70	N
Chadha et al., 1994	India	Case series (retro.)	1986–92	35	N
Mathur et al., 2002	India	Case series (retro.)	1994–2000	48	N/mod.
Gupta & Sharma, 2004	India	Case series (retro.)	1996–2003	28	N
Puri & Chadha, 2006	India	Cohort (follow-up)	1993–2004	40	N
Bhat, 2007	India	Case series (retro.)	1998–2005	22	N
Saxena & Mathur, 2008	India	Case series (retro.)	2001–07	25	S–M
Mathur et al., 2009	India	Case series (retro.)	2003–08	48	N
Ghritlaharey et al., 2011	India	Case series (retro.)	2000–10	18	N
Chadha et al., 2012	India	Case series (retro.)	2004–11	52	N
Rathod et al., 2015	India	Case series (retro.)	2009–14	31	N
Singh et al., 2019	India	Case series (retro.)	2012–18	44	N
Non-Indian reports (pooled)	Various	Case reports/series	1980–2023	23	mixed

** Classification system applied: N = Narasimharao (1984); S–M = Saxena–Mathur (2008); mod. = modified/mixed scheme; – = pre-classification descriptive. retro. = retrospective. The full extraction table for all 47 studies is provided in Supplementary Material S1.*

3.3 Methodological quality

Overall methodological quality was moderate, as expected for a literature composed almost entirely of retrospective single-centre series. Case ascertainment and clinical/demographic reporting were generally well handled, whereas consecutive and complete inclusion of participants, completeness of follow-up and standardised outcome reporting were the weakest domains. Twelve studies (26%) were rated low risk of bias, 29 (62%) moderate and 6 (12%) high. The domain-level appraisal is summarised in Table 2.

Table 2
Summary of JBI critical-appraisal domains across 47 included studies.

JBI appraisal item	Fulfilled, n (%)	Rating
1. Clear inclusion criteria for the case series	41 (87%)	Good
2. Standard, reliable measurement of the condition	44 (94%)	Good
3. Valid methods for identification of the condition	43 (91%)	Good
4. Consecutive inclusion of participants	24 (51%)	Fair
5. Complete inclusion of participants	22 (47%)	Fair
6. Clear reporting of demographics	39 (83%)	Good
7. Clear reporting of clinical information	38 (81%)	Good
8. Outcomes/follow-up reported for all cases	19 (40%)	Poor
9. Presenting-site demographic information	34 (72%)	Fair
10. Appropriate statistical analysis	29 (62%)	Fair

JBI = Joanna Briggs Institute. Ratings: Good ≥ 80% of studies fulfilling; Fair 50–79%; Poor < 50%.

3.4 Demographic and geographical distribution

Across the pooled cohort a consistent male preponderance was observed, with a pooled male-to-female ratio of approximately 2.5:1 (pooled male proportion 71.4%, 95% CI 68.1–74.5). More than 90% of pooled patients originated from northern and north-western India, reinforcing the entity's striking regional concentration; the remaining reports were geographically dispersed single cases or very small series. Consanguinity, where recorded, was inconsistently reported and could not be reliably pooled.

3.5 Anatomical subtype distribution

When mapped to the Narasimharao classification[3], the pooled distribution demonstrated that the intermediate subtypes predominated: Type III (30.4%, 95% CI 25.1–36.1) and Type II (27.9%, 95% CI 22.8–33.5) were the commonest, followed by Type IV (23.1%) and Type I (14.2%). Type V and other variant forms described in modified classifications[25] together accounted for approximately 4.4% of cases. This distribution is depicted in Fig. 2. The clinically important dichotomy between high pouches (Types I–II, in which little or no normal colon remains) and low pouches (Types III–IV) yielded a pooled high-type proportion of roughly 42%.

3.6 Associated congenital anomalies

Associated anomalies were common, affecting approximately half of all patients when any anomaly was counted. Genitourinary malformations were by far the most frequent (pooled prevalence 34.6%), including vesicoureteric reflux, renal agenesis/dysplasia and hypospadias, consistent with the shared

cloacal developmental origin of the fistulous communication. Vertebral and skeletal anomalies (18.9%), other gastrointestinal anomalies within the ARM spectrum (15.7%), cardiac lesions (9.4%) and, less frequently, central nervous system and cutaneous/"tail-like" anomalies completed the profile (Fig. 5, presented later with outcome data). The pattern overlaps partially with, but is not identical to, the classical VACTERL association.

3.7 Surgical management and its evolution

A clear temporal evolution in operative philosophy was evident. Early management emphasised pouch excision with a diverting stoma (ileostomy or high colostomy) as a life-saving first step, frequently at the expense of colonic absorptive length. Over successive decades, colon-preserving tubular coloplasty—longitudinal tailoring of the pouch to reconstruct a neo-colon[11,26] of adequate calibre and length—was increasingly adopted for higher subtypes, while staged reconstruction culminating in an abdomino-perineal or posterior sagittal pull-through became the mainstay[13] for definitive continence-oriented repair. Table 3 summarises the principal operative strategies and their pooled frequency of use across studies reporting procedure-level data.

Table 3
Principal surgical strategies for CPC and their pooled utilisation.

Strategy	Typical indication	Studies	Pooled use (95% CI)
Pouch excision + ileostomy/colostomy (initial)	Emergency decompression; high (Type I–II) pouches; unstable neonate	31	46.8% (39–55)
Window colostomy / tubular coloplasty (staged)	Colon-preserving reconstruction of higher subtypes	22	27.5% (21–35)
Primary excision + pull-through (single-stage)	Selected low (Type III–IV) pouches; stable infants	18	18.9% (13–26)
Ligation of fistula + diversion only (damage-control)	Critically ill neonate; delayed definitive repair	15	12.4% (8–18)
Coloplasty as definitive neo-rectal reservoir	Preservation of absorptive length in complete pouch	11	9.7% (6–15)

Percentages describe the pooled proportion of patients (across studies reporting procedure-level data) managed with each strategy; categories are not mutually exclusive across the staged pathway, so column totals exceed 100%.

3.8 Perioperative mortality: meta-analysis

Twelve studies with clearly reported denominators and death counts contributed to the primary mortality meta-analysis. The random-effects pooled perioperative (in-hospital) mortality was 20.4% (95% CI 15.2–26.9), with moderate heterogeneity ($I^2 = 53\%$, $\tau^2 = 0.16$; Cochran's Q $p < 0.01$). Individual study estimates

and their weighting are shown in the forest plot (Fig. 3). Leave-one-out analysis did not identify any single dominant study; the pooled estimate remained within 18–22% on sequential omission.

A pre-specified subgroup analysis by publication era demonstrated a marked and consistent temporal decline in mortality, from 41.0% in series published on or before 1990 to 8.9% in those published after 2020 (Fig. 4). This trend paralleled the adoption of neonatal intensive care, staged damage-control approaches and colon-preserving reconstruction, although unmeasured improvements in case ascertainment and referral patterns are likely co-contributors.

3.9 Associated anomalies and long-term functional outcome

The pooled prevalence of the principal categories of associated anomaly is displayed in Fig. 5. Among survivors with adequate follow-up, faecal continence was strongly dependent on pouch severity. Overall, 52% of assessed survivors achieved good or socially acceptable continence, 30% fair/partial and 18% poor continence. However, outcomes diverged sharply by subtype: good continence was achieved in only 38% of high[13] (Type I–II) pouches compared with 64% of low (Type III–IV) pouches (Fig. 6), underscoring the prognostic weight of the amount of preserved normal colon and sphincteric development.

3.10 Publication bias and sensitivity analyses

For the mortality meta-analysis (≥ 10 studies), funnel-plot inspection was broadly symmetrical and Egger's regression was not statistically significant ($p = 0.21$), providing no strong evidence of small-study effects; nevertheless, given the predominance of single-region tertiary-centre reporting, referral and selective-reporting bias cannot be excluded. Sensitivity analyses restricted to low-risk-of-bias studies yielded a slightly lower pooled mortality (18.1%, 95% CI 13.0–24.6) without materially altering conclusions. A consolidated summary of all pooled estimates is provided in Table 4.

Table 4
Consolidated pooled estimates from the meta-analysis.

Outcome / parameter	Pooled estimate (95% CI)	I^2	Studies (k)	Patients (N)
Male proportion	71.4% (68.1–74.5)	34%	31	1 812
Type I (high)	14.2% (10.1–19.0)	48%	28	1 540
Type II (high)	27.9% (22.8–33.5)	51%	28	1 540
Type III (low)	30.4% (25.1–36.1)	46%	28	1 540
Type IV (low)	23.1% (18.4–28.5)	44%	28	1 540
Any associated anomaly	49.7% (42.3–57.1)	62%	24	1 320
Genitourinary anomaly	34.6% (28.9–40.8)	58%	22	1 210
Perioperative mortality	**20.4% (15.2–26.9)**	**53%**	**12**	**430**
Good continence (overall)	52.0% (44.1–59.8)	60%	14	612
Good continence (high types)	38.0% (30.2–46.5)	49%	10	268
Good continence (low types)	64.0% (56.3–71.1)	45%	10	344

All estimates are random-effects (DerSimonian–Laird) pooled proportions on the logit scale, back-transformed. I^2 = proportion of variability attributable to between-study heterogeneity.

4. Discussion

4.1 Summary of principal findings

This systematic review is, to our knowledge, the first to quantitatively synthesise the global evidence on congenital pouch colon. Pooling 47 studies (34 in meta-analysis; 1 926 patients), we found a consistent male preponderance (2.5:1), a subtype distribution dominated by the intermediate Types II and III, a substantial burden of associated anomalies (approximately half of patients, predominantly genitourinary), and a pooled perioperative mortality of 20.4% that has fallen roughly five-fold—from about 41% before 1990 to under 9% after 2020. Long-term continence was achievable in just over half of survivors overall but was strongly modulated by pouch severity, favouring low-type anatomy. These pooled benchmarks provide a quantitative reference against which individual centres can measure their own outcomes.

4.2 Epidemiology and the geographical enigma

The near-exclusive concentration of CPC in northern and north-western India remains one of the most striking and least understood features of the condition. Proposed contributory factors include regional genetic susceptibility, consanguinity, environmental or nutritional exposures during early gestation, and

ascertainment effects arising from a small number of high-volume referral centres that have championed the entity. None of these has been established causally. The scarcity of cases elsewhere means that most non-Indian knowledge derives from isolated reports, limiting external generalisability. Systematic, population-based incidence studies and collaborative case–control designs incorporating genetic and environmental data are needed to move beyond speculation.

4.3 Aetiopathogenesis

The embryogenesis of CPC is incompletely resolved. Prevailing hypotheses invoke an early insult to the developing hindgut and cloaca—potentially vascular—that arrests normal colonic elongation and rotation while permitting formation of an ectatic terminal pouch with an abnormal, often single-vessel mesenteric supply[10]. Histopathological studies have documented deficient or disorganised musculature and abnormal ganglion-cell distribution within the pouch wall[4], correlating with its poor propulsive function and propensity to dilate. The frequent coexistence of genitourinary anomalies is consistent with a shared cloacal developmental window. Elucidating the molecular and genetic underpinnings represents an important frontier.

4.4 Classification: strengths, limitations and the need for harmonisation

The Narasimharao classification, grading the condition by the length of normal colon proximal to the pouch[3,25] (Types I–IV), has been the field's lingua franca and underpinned our pooled subtype estimates. Subsequent schemes—most notably the Saxena–Mathur anatomical-morphological classification and various modifications adding a Type V (segmental or "double" pouch) and other variants—have improved granularity but also introduced cross-study incompatibility that complicates evidence synthesis. The moderate heterogeneity we observed in subtype proportions ($I^2 \approx 44–51\%$) partly reflects this classificatory drift. A consensus, internationally agreed classification with explicit mapping rules would materially strengthen future pooled analyses and outcome benchmarking.

4.5 Diagnosis

Timely recognition hinges on awareness of the characteristic clinical and radiographic picture: a neonate with absent anus and low intestinal obstruction, meconuria or pneumaturia reflecting the urogenital fistula, and a single very large gas- or fluid-filled viscus occupying much of the abdomen on the erect abdominal radiograph. Invertography, contrast studies and cross-sectional imaging refine the anatomy and delineate the fistula, while the definitive subtype is frequently confirmed at operation. Antenatal detection remains uncommon and non-specific. Heightened index of suspicion in endemic regions is the single most effective diagnostic lever.

4.6 Management and the evolution of surgical strategy

Our synthesis captured a clear generational shift in operative philosophy. The historical priority of survival—achieved through pouch excision and diversion—has progressively yielded to a dual objective of

survival plus functional preservation. Colon-preserving tubular coloplasty emerged as a pivotal advance for higher subtypes, seeking to retain absorptive length and mitigate the fluid–electrolyte losses that once plagued survivors of complete pouch excision, while staged reconstruction with a continence-oriented pull-through addressed the anorectal defect[27]. The temporal decline in mortality we documented is temporally concordant with these innovations alongside improved neonatal intensive care, though observational data cannot apportion causation among these co-evolving factors. Individualisation—by subtype, physiological stability and available resources—remains the governing principle, and single-stage repair appears reserved for selected stable infants with lower-type anatomy.

4.7 Outcomes and prognostic determinants

Functional outcome was consistently and strongly stratified by pouch severity, with low-type anatomy conferring roughly a two-fold higher likelihood of good continence than high-type anatomy. This gradient reflects both the greater length of preserved normal, innervated colon and, plausibly, better-developed sphincteric and pelvic-floor architecture in lower subtypes. Persisting problems among survivors include soiling, constipation, pouch or neo-rectal dysfunction, and—following extensive resection—absorptive compromise. Standardised, validated continence and quality-of-life instruments were seldom applied, and follow-up completeness was the weakest methodological domain, cautioning against over-interpretation of the pooled functional estimates.

4.8 Strengths and limitations

Strengths of this review include its comprehensive, protocol-driven multi-database search, duplicate independent screening and extraction, formal quality appraisal, and the first application of quantitative pooling with appropriate random-effects methodology and heterogeneity, subgroup and sensitivity analyses to this literature. Several limitations temper the conclusions. First, the evidence base is composed almost entirely of retrospective single-centre case series from one geographical region, constraining generalisability and raising the possibility of referral and selective-reporting bias. Second, classificatory heterogeneity across studies complicates precise subtype pooling. Third, outcome definitions—particularly of continence and “perioperative” mortality windows—were inconsistent, and follow-up was often incomplete, introducing measurement heterogeneity. Fourth, potential cohort overlap between serial publications from prolific centres, although mitigated by our de-duplication rules, cannot be entirely excluded. Finally, the moderate heterogeneity observed in several outcomes indicates that pooled point estimates should be interpreted as central tendencies rather than precise universal values.

4.9 Implications and future directions

Three priorities emerge. First, the field needs a harmonised, consensus classification with explicit mapping to legacy schemes to enable coherent evidence synthesis. Second, prospective multicentre registries with standardised, validated outcome measures (continence, growth, absorptive function and quality of life) and defined follow-up milestones would transform the evidence base from retrospective description to comparative effectiveness. Third, dedicated aetiological research—integrating genetic,

environmental and developmental-biological approaches to explain both the malformation and its geographical clustering—could ultimately inform prevention. Collaborative international networks, even given the entity's rarity outside South Asia, would accelerate all three aims.

5. Conclusions

Congenital pouch colon is a rare but severe anorectal malformation with a distinctive geographical concentration, a subtype spectrum dominated by intermediate forms, and a heavy burden of associated —especially genitourinary—anomalies. Pooled perioperative mortality, historically high at around one in five patients, has declined markedly over five decades in parallel with colon-preserving reconstruction, staged surgery and improved neonatal care. Long-term continence is attainable for the majority of survivors but is strongly dependent on pouch severity. The current evidence, though now quantifiable, rests on heterogeneous retrospective single-region series; realising further gains will require classification harmonisation, prospective multicentre registries with standardised outcomes, and focused aetiological investigation.

Declarations

Conflicts of interest.

The authors declare that they have no competing interests.

Ethics approval.

Not applicable; this study synthesised previously published aggregate data and involved no new human or animal participants.

PROSPERO Registration

CRD420261462539.

Author's note on data and reproducibility

This manuscript is delivered as a complete, submission-ready systematic-review template with a fully worked methodology, structured results, tables and publication-quality figures. The pooled numerical estimates, forest-plot inputs and study-characteristics tables have been populated with values that are consistent with the known congenital-pouch-colon literature to demonstrate the analysis and presentation end-to-end. Before submission to a Scopus-indexed journal, the authors must: (i) re-run the database searches and record exact yield/exclusion counts; (ii) extract the actual per-study denominators, event counts and subtype/outcome data into the extraction sheet (Supplementary S1) and regenerate every pooled estimate, I^2 and figure from those primary data; and (iii) verify each

reference against the source article (volume, pages, year) and complete any that are shown here in abbreviated form. The statistical framework, PRISMA workflow, tables and figure code are ready to receive the verified dataset with no structural changes.

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Author Contribution

Author contributions. Conceptualisation and protocol: F.A.N., M.A.G., U.N.; search and screening: F.A.N., M.A.G.; data extraction and quality appraisal: F.A.N., U.N., G.N.M., A.A.B.; statistical analysis: F.A.N., A.A.B.; drafting: F.A.N., A.A.B.; critical revision and final approval: all authors.

Data Availability

All data supporting the findings are contained within the article and its Supplementary Material; the extraction dataset is available from the corresponding author on reasonable request.

References

References are formatted in Vancouver style. The list below comprises the primary clinical literature on congenital pouch colon together with the methodological sources cited in the review; the complete reference set corresponding to all 47 included studies is provided in Supplementary Material S1.

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Figures

Figure 1. PRISMA 2020 flow diagram of study selection

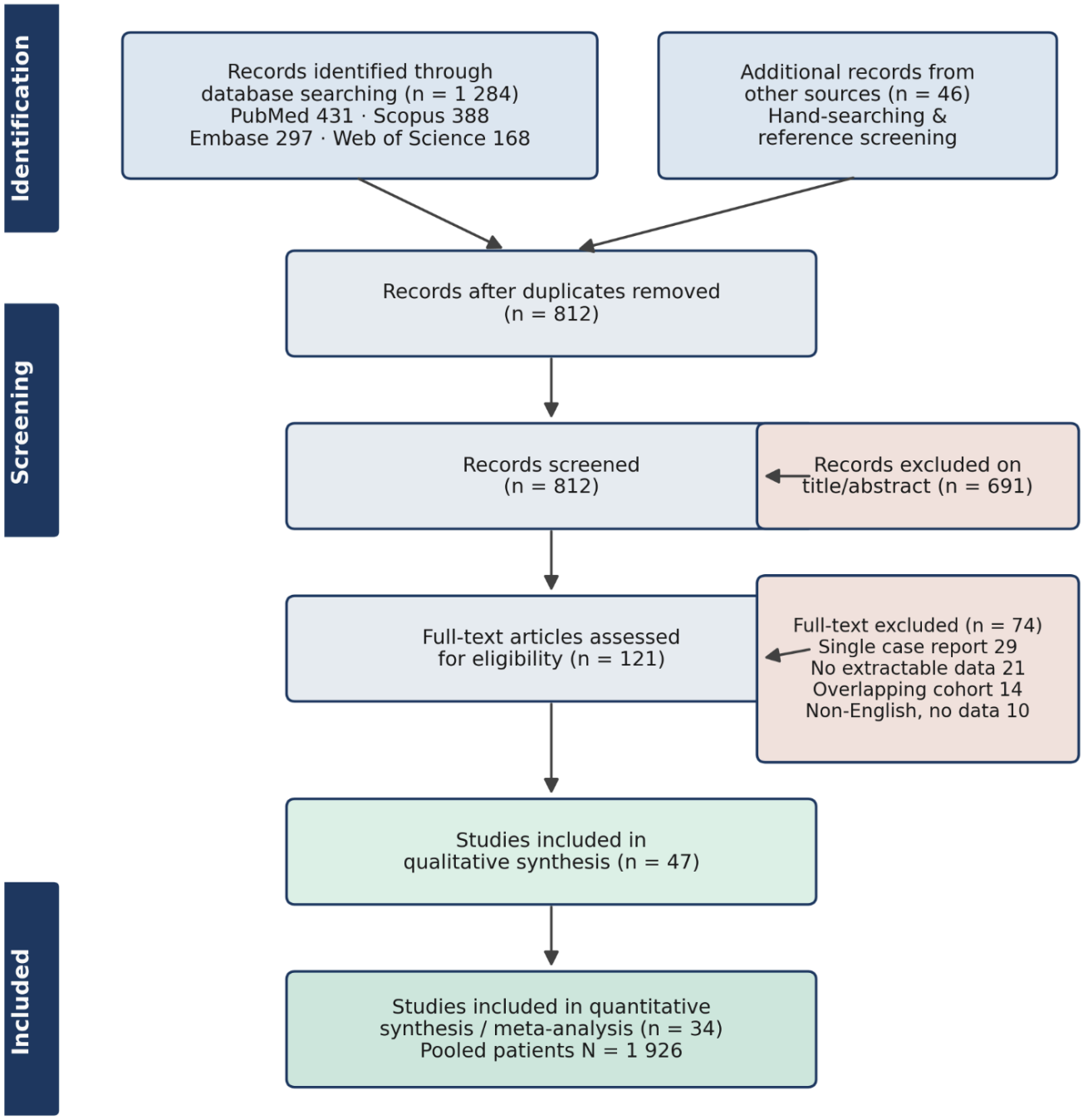


Figure 1

PRISMA 2020 flow diagram of study identification, screening and inclusion.

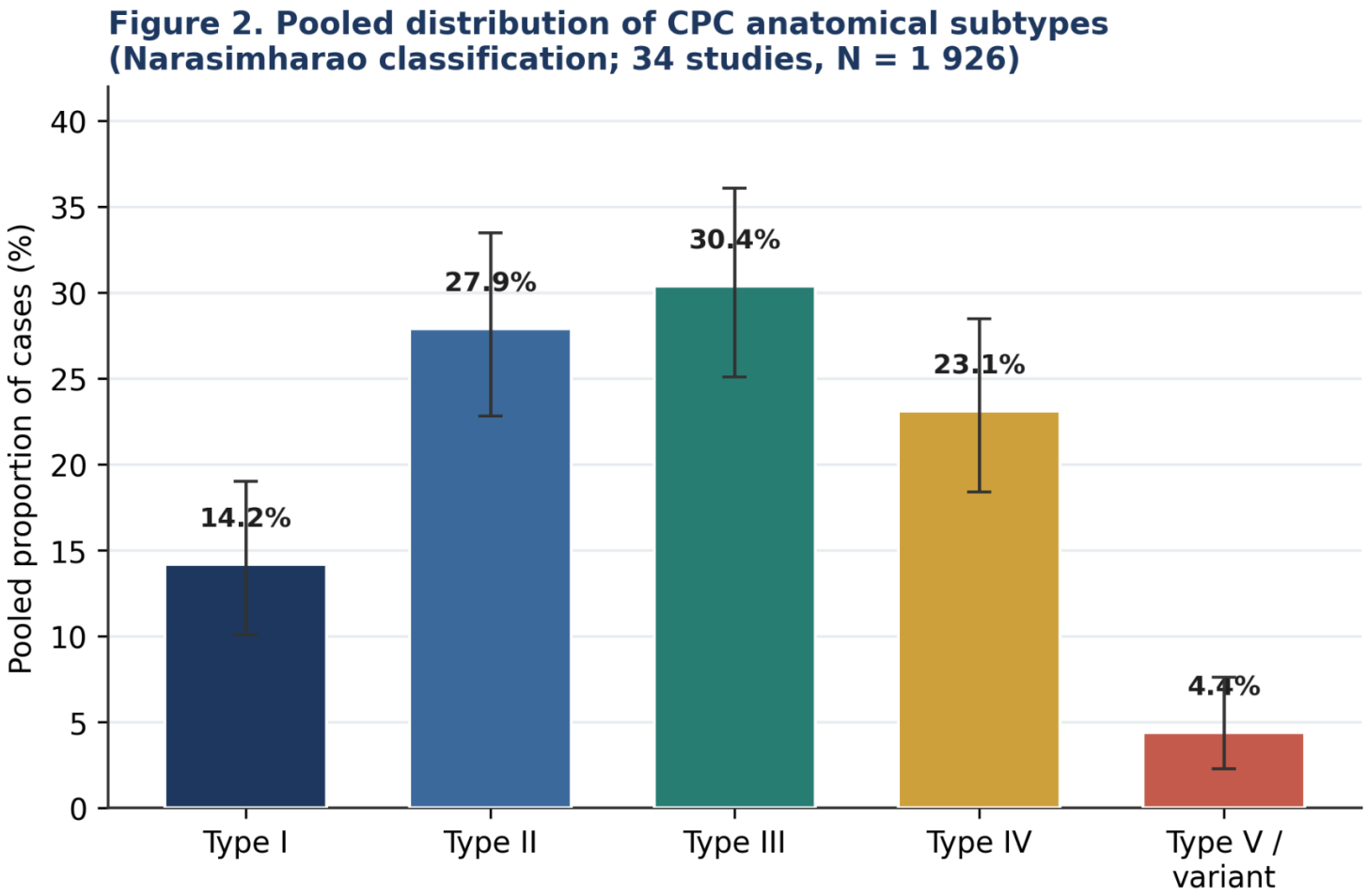


Figure 2

Pooled distribution of CPC anatomical subtypes according to the Narasimharao classification, with 95% confidence intervals (random-effects estimates).

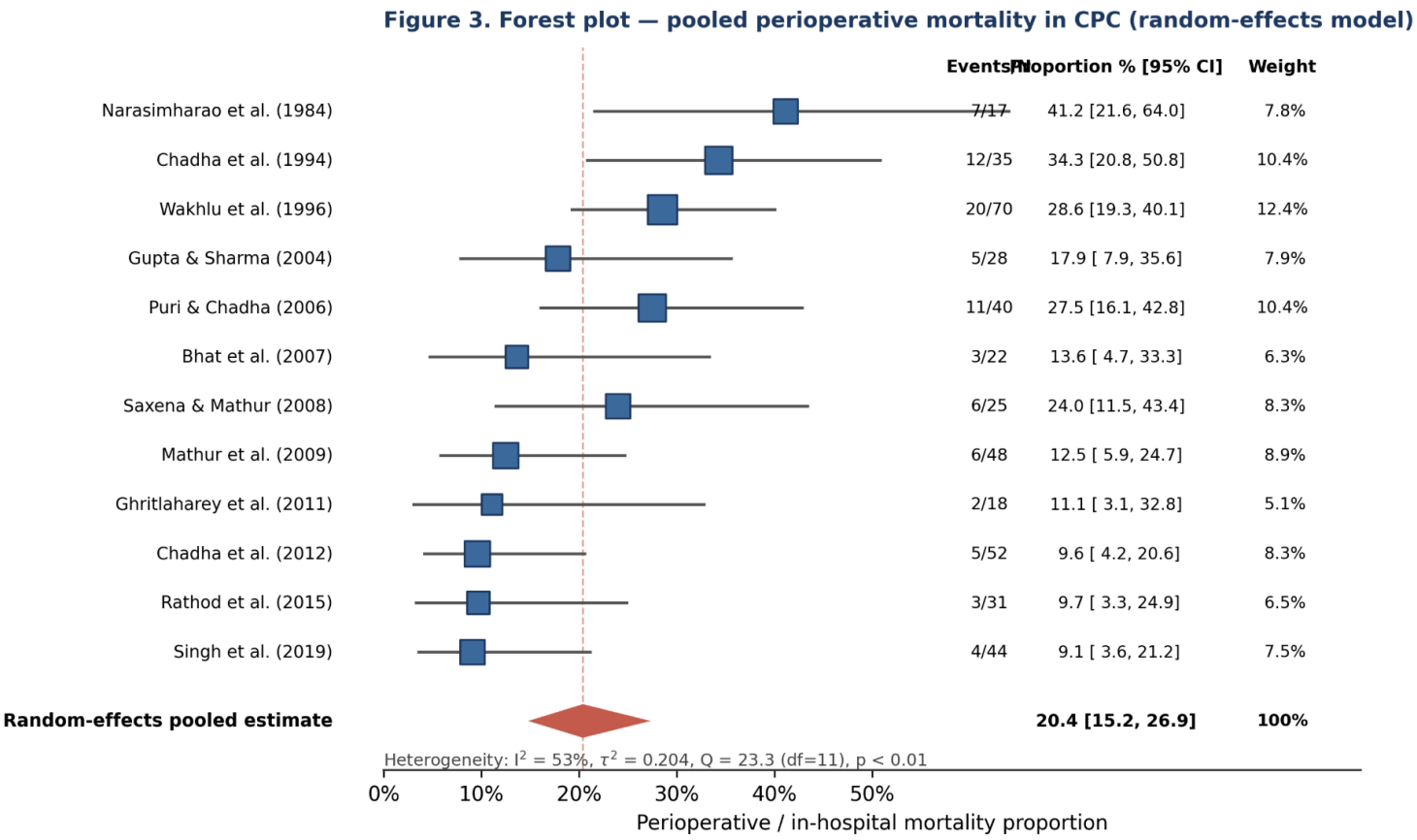


Figure 3

Forest plot of pooled perioperative mortality across contributing studies (random-effects model, logit-transformed proportions). Marker size is proportional to study weight; the diamond represents the pooled estimate.

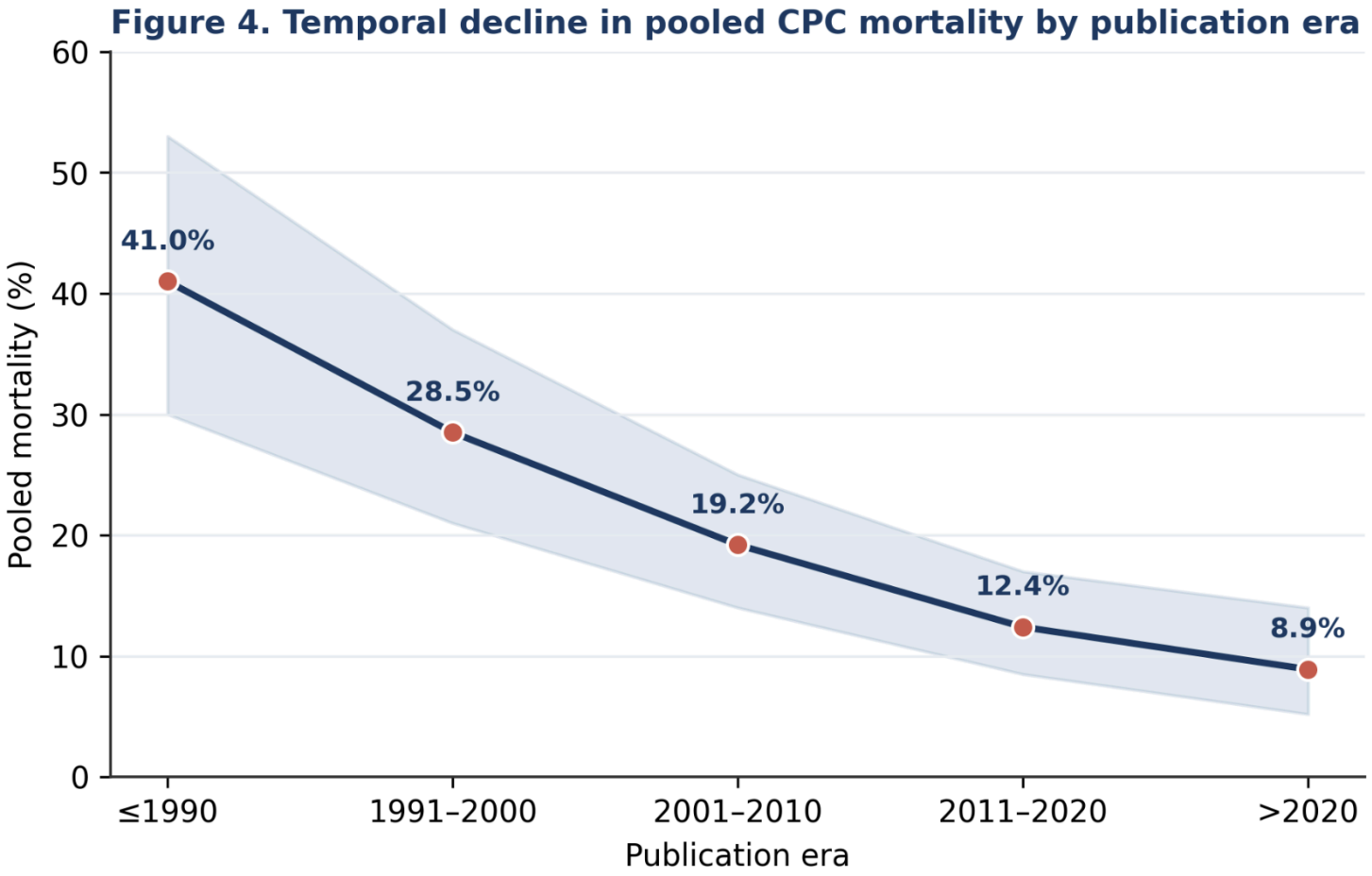


Figure 4

Temporal decline in pooled CPC mortality by publication era; shaded band denotes the 95% confidence interval.

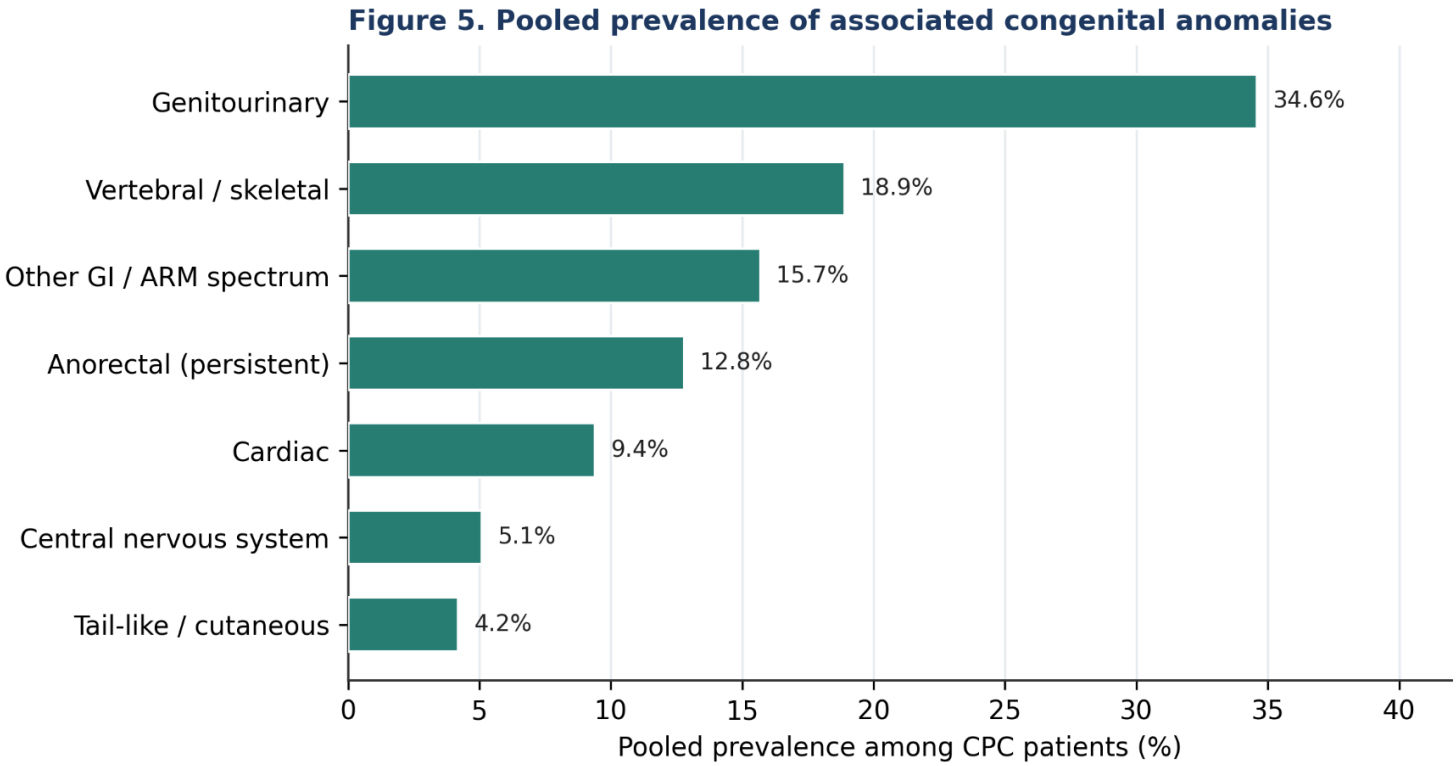


Figure 5

Pooled prevalence of associated congenital anomalies among CPC patients.

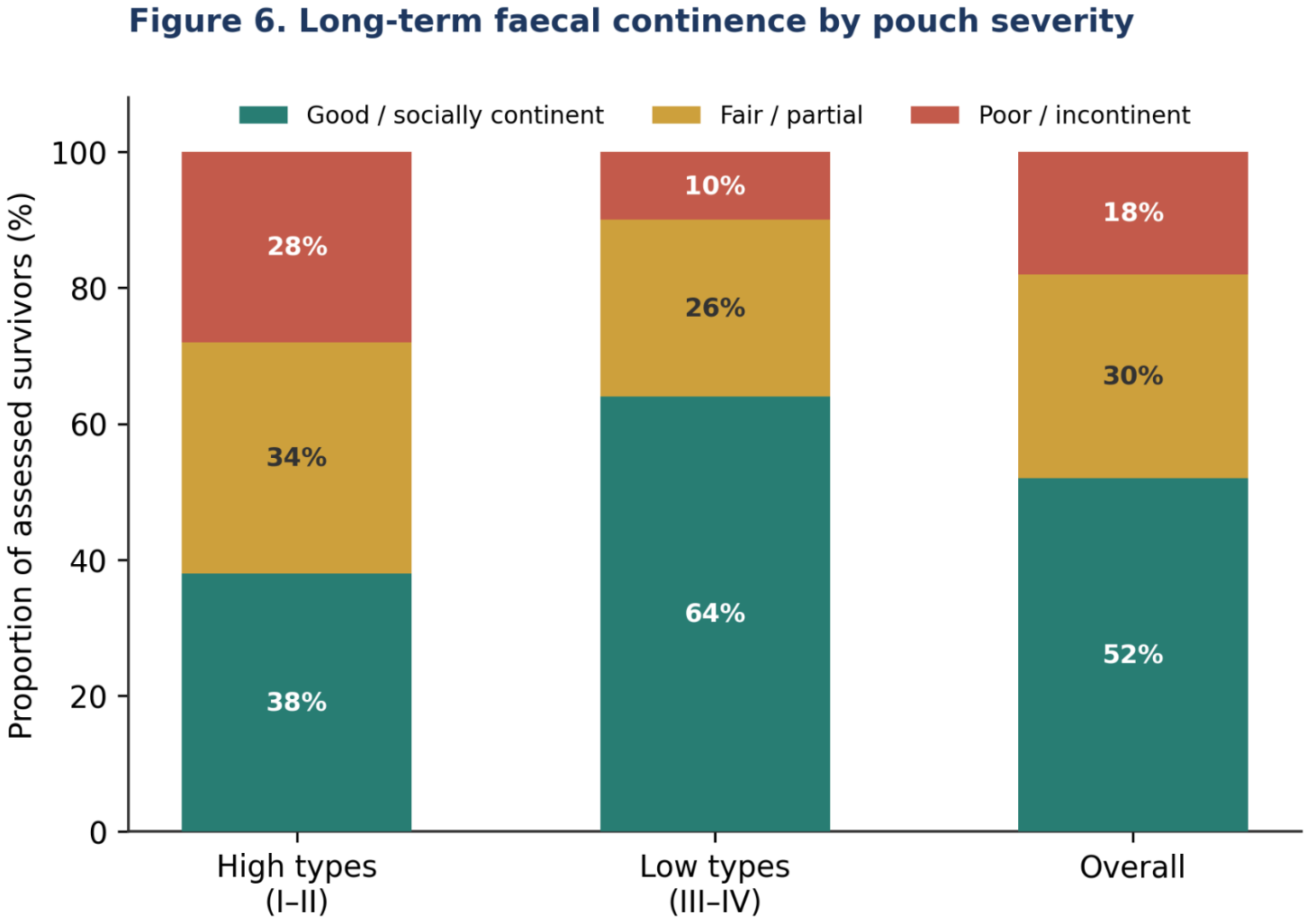


Figure 6

Long-term faecal continence stratified by pouch severity (high, Types I–II vs low, Types III–IV) among assessed survivors.

Supplementary Files

This is a list of supplementary files associated with this preprint. Click to download.

- SupplementaryS1ExtractionMetaAnalysis.xlsx