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Quantifying the impact of congenital cardiac co-morbidities on anorectal malformations

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ABSTRACT

Aims of the study: Anorectal malformations (ARM) are frequently associated with congenital heart defects (CHD) (prevalence 10–40 %). Cardiac ARM patients, particularly those requiring surgery, tend to experience delays in definitive anorectal reconstruction whilst awaiting favourable cardiac status. We sought to quantify these delays and evaluate CHD's impact on surgical complications.

Method: ARM patients treated at a single cardiac centre were identified through a prospectively maintained database. A consultant cardiologist reviewed all patient cardiac studies and divided them into either a *cardiac* or *non-cardiac* group based on their functional status ± need for cardiac surgery. Demographics, ARM classification, surgical history, and complications were analysed using SPSS (IBMv31). Mann-Whitney-U and Chi-squared tests evaluated continuous and categorical variables, respectively. Complication counts were stratified by surgical time-point, with odds ratios calculated ($p < 0.05$ = significant).

Results: 62 ARM patients were identified over 7 years (2018–2024); *cardiac* (n = 19) and *non-cardiac* (n = 43). Cardiac patients underwent definitive anorectal reconstruction when significantly older (396 vs. 175 days, $p < 0.001$), even after adjusting for birthweight and gestation ($p = 0.002$). One or more complication occurred in 47 % of cardiac patients versus 27 % of non-cardiac patients (OR 2.33, 95 % CI 0.76–7.13, $p = 0.136$), an insignificant difference. Cardiac status was however a statistically significant predictor of total complication count (Wald $\chi^2 = 4.049$, $p = 0.044$), with Poisson regression models showing cardiac patients had 2.11 times the rate of complications compared to non-cardiac patients (Incidence Rate Ratio/IRR = 2.11, 95 % CI: 1.23–4.37). Each additional day from stoma formation to closure increased complication likelihood by 0.4 % (OR 1.004, 95 % CI 1.000–1.008, $p = 0.029$).

Conclusions: Cardiac ARM patients undergo definitive reconstruction significantly laterwith a greater proportion suffering complications compared with their non-cardiac counterparts. Quantifying these delays and risks provides clinicians with data to guide prognostic counselling, proactively address psychosocial burdens, and refine multidisciplinary protocols shaping the ARM-CHD patients' surgical journey. Further work is crucial to disentangling relationships between CHD severity, circulatory physiology, and clinician imposed surgical delay to better define their exact contributions to risk.

1. Introduction

Anorectal malformations (ARM) are rare congenital anomalies of the distal gastrointestinal tract, estimated to occur globally in 1 in 4000 live births [1,2]. Over 60 % of ARM patients present with associated anomalies, with cardiac defects amongst the most frequently observed [2,3]. The reported prevalence of congenital heart defects (CHD) in ARM varies between 10 % and 40 %, with severity appearing to correlate with complexity of ARM phenotype in some studies [4,5]. While atrial and ventricular septal defects predominate and may be managed conservatively, a significant proportion of patients have complex cyanotic cardiac defects requiring surgical intervention within the first month of life [6]. Reconstructive surgery in anorectal malformations typically involves 3-stage surgery with a diverting ostomy, surgical construction of the anomaly and subsequent stoma closure. Despite anatomical correction, studies show 50–70 % of patients report

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persistent bowel dysfunction, including constipation and incontinence conferring high disease and healthcare system burden [2,7]. These outcomes are closely tied to anatomical features and the timing of surgical intervention [8].

Emerging evidence suggests that timing of anorectal reconstruction is a critical determinant of long-term outcome [8]. Early PSARP, performed within the first two to three months of life, has been associated with reduced stoma-related complications, improved continence, and fewer surgical stages [8–10]. Delays beyond 6–9 months have been correlated with increased need for bowel management and lower functional scores in some studies [8,11]. These associations underscore the importance of minimising unnecessary postponement of repair. Delayed reconstruction also imposes additional burden on families and healthcare systems. Prolonged stoma care is associated with increased re-admissions, surgical revisions, and infections, while also affecting quality of life, caregiver stress, and psychosocial development [2,8,12]. These cumulative risks may be magnified in patients already experiencing the clinical complexity and monitoring demands of congenital cardiac disease. Anecdotally, in clinical practice we have observed that patients with a CHD diagnosis often undergo reconstructive surgery at an older age. It remains unclear whether this surgical delay is protective against or contributes to post-operative morbidity, or whether it is merely a surrogate marker of cardiac fragility. Children with CHD, particularly those with impaired perfusion, cyanosis, or cardiopulmonary instability, may face increased peri-operative risk when undergoing non-cardiac procedures, with higher rates of anaesthetic morbidity, reintubation, and post-operative complications [13–15] This may partially explain why definitive ARM reconstruction is frequently postponed until cardiac optimisation is achieved [3,12].

Despite its clinical relevance, no prior studies have quantified the surgical delays experienced by ARM patients with CHD or evaluated the impact of cardiac status on ARM post-operative outcomes. This study aims to address this evidence gap through a retrospective cohort analysis of ARM patients at a specialist, tertiary paediatric surgical and cardiac centre. The objectives were to: (1) quantify the delay in definitive anorectal reconstruction in patients with CHD; and (2) assess whether CHD diagnosis, independently or in interaction with surgical delay, is associated with increased post-operative complications.

2. Methods

2.1. Study design and data collection

This was a retrospective cohort study conducted at a tertiary referral centre for paediatric cardiac and general surgery (Evelina London Children's Hospital, United Kingdom). All patients diagnosed with anorectal malformations (ARM) between July 2018 and July 2024 were identified via a prospectively maintained institutional database. Electronic medical records (EPIC) and the London Care Record were reviewed retrospectively by two independent investigators (SS, HT).

Data were extracted into a structured spreadsheet, incorporating the following pre-defined variables; Demographics: sex, gestational age, birthweight, age at ARM diagnosis, ARM Classification: Krickenbeck subtype delineated on loopogram imaging/intra-operatively, Associated Anomalies: VACTERL components (vertebral/spinal, anorectal, cardiac, tracheoesophageal, renal, and limb), genetic diagnoses, Cardiac Data: congenital heart disease (CHD) presence and type, ECHO findings, cardiac surgery status; Surgical Timelines: patient age (in days) at three key time points: at stoma formation, definitive anorectal reconstruction, and stoma closure. All post-operative complications were categorised into binary (0 or ≥1) and ordinal fields (count incidence) and stratified by surgical stage into 4 'time periods': *(1) between stoma formation and definitive reconstruction, (2) between definitive reconstruction and stoma closure, (3) at stoma closure, and (4) late complications following surgical repair.* A subset of patients underwent primary anoplasty and did not require ostomy formation.

Patients were excluded if deceased, or if follow-up data were incomplete due to transfer of care prior to definitive reconstruction. Duration of follow-up was variable and dependent on patient age and time since initial presentation but extended to a minimum of the data cut-off point in all cases.

2.2. Patient grouping criteria

Patients were stratified into two main cohorts based on ECHO-confirmed cardiac anatomy and status as reviewed by a consultant paediatric cardiologist:

- **Cardiac group:** patients with left-to-right shunts undergoing surgery in infancy, duct-dependent systemic circulation requiring biventricular neonatal repair, duct-dependent pulmonary circulation requiring biventricular staged-surgery and other complex anomalies requiring surgery in the early neonatal period or infancy.
- **Non-cardiac group:** patients with normal ECHO findings, or with minor CHD (e.g., self-resolving patent foramen ovale or small atrial septal defects) not requiring intervention.

Cardiac surgery status was recorded separately but was not used to define cohort allocation, thereby allowing for isolated analysis of the effect of cardiac diagnosis itself, independent of surgical correction.

2.3. Statistical analysis

Data were analysed using IBM SPSS Statistics v3. Continuous variables were summarised as medians with interquartile ranges (IQR) and compared using the Mann–Whitney U test. Categorical variables were compared using the Chi-squared test or Fisher's exact test, as appropriate. Binary logistic regression was used to evaluate the association between CHD status and the presence of ≥1 postoperative complication, with odds ratios (OR) and 95 % confidence intervals (CI) reported. To assess the combined and independent effects of CHD status and surgical delay, Poisson regression modelling was performed using complication count as the outcome. A p-value <0.05 was considered statistically significant.

2.4. Ethical considerations

This study was conducted in accordance with institutional guidance for audit and service evaluation. As it involved the secondary use of routinely collected clinical data without intervention or patient identifiers, formal research ethics approval and individual consent were not required.

3. Results

3.1. Demographics

A total of 62 patients with anorectal malformations (ARM) were identified between July 2018 and July 2024.

Table 1
Results summary of entire cohort of patients.

DEMOGRAPHICS		Cardiac	Non-cardiac	Total	Significance (p value)
Total number (n)		**19 (31 %)**	**43 (69 %)**	**62**	
Gestation (median, weeks)		38 (25–42)	38 (32–41)	38	0.269
Birth weight (mean, grams)		2659	2886	2814	0.351
Sex (female (n): Male (n))		M15:F4	M30:F13	M45:F17	0.65
VACTERL > 3 (n)		15 (79 %)	15 (35 %)	30 (48 %)	**<0.001***
Cardiac surgery performed (n)		18 (95 %)	0 (0 %)	18 (29 %)	N/A
Primary anoplasty performed (no stoma)		1 (5 %)	7 (16 %)	8 (13 %)	0.06
ARM anomaly KRICKENBECK classification (n,%)					
Female	Perineal fistula	2 (11 %)	7 (16 %)	9 (15 %)	0.553
	Rectovestibular fistula	2 (11 %)	6 (14 %)	8 (13 %)	0.711
	Anal stenosis	0 (0 %)	0 (0 %)	0 (0 %)	N/A
Male	Rectobulbar fistula	7 (37 %)	9 (21 %)	16 (26 %)	0.489
	Perineal fistula	4 (21 %)	10 (23 %)	14 (23 %)	0.848
	Rectoprostatic fistula	3 (16 %)	4 (9 %)	7 (10 %)	0.279
	Imperforate anus no fistula	1 (5 %)	4 (9 %)	5 (8 %)	0.590
	Rectobladder fistula	0 (0 %)	2 (5 %)	2 (3 %)	N/A
	Anal stenosis	0 (0 %)	1 (2 %)	1 (2 %)	N/A
Surgical timeline (median days, range)					
Age at stoma formation		2 (1–356)	1 (0–301)	–	0.205
Age at definitive reconstruction		396 (186–1072)	175 (1–557)	228	**<0.001***
Age at stoma closure		554 (269–1312)	265 (73–1656)	329	**<0.001***
Days between reconstruction and stoma closure		121 (50–938)	75 (30–1603)	–	**0.048***
Complications					
Total patients with ≥ 1 complication (n)		**9 (47 %)**	**12 (27 %)**	**21 (34 %)**	**0.136**
Total incidence of complication		**14**	**15**	**29**	**0.041***
1	**Time period 1 (total)**	**4 (31 %)**	**5 (29 %)**	**9 (31 %)**	
Between stoma formation and definitive reconstruction	⇒ unplanned emergency laparotomy	3 (23 %)	2 (12 %)	5 (17 %)	
	⇒ stoma site infection/dehiscence	0 (0 %)	2 (12 %)	2 (7 %)	
	⇒ elective stoma revision	1 (8 %)	1 (6 %)	2 (7 %)	
2	**Time period 2 (total)**	**4 (31 %)**	**5 (29 %)**	**9 (31 %)**	
Between definitive reconstruction and stoma closure	⇒ Re-do anoplasty (stricture)	1 (8 %)	0 (0 %)	1 (3 %)	
	⇒ Re-do anoplasty (prolapse/retraction)	2 (15 %)	3 (18 %)	5 (17 %)	
	⇒ perineal wound dehiscence	1 (8 %)	1 (6 %)	2 (7 %)	
	⇒ sepsis, medically managed	0 (0 %)	1 (6 %)	1 (3 %)	
3	**Time period 3 (total)**	**4 (23 %)**	**1 (6 %)**	**5 (17 %)**	
At/Post stoma closure	⇒ anastomotic leak	0 (0 %)	1 (6 %)	1 (3 %)	
	⇒ SSI/Wound dehiscence	2 (15 %)	0 (0 %)	2 (7 %)	
	⇒ sepsis, medically managed	2 (15 %)	0 (0 %)	2 (7 %)	
4	**Time period 4 (total)**	**2 (15 %)**	**4 (24 %)**	**6 (21 %)**	
Late complications	⇒ neoanus stenosis (revised)	1 (8 %)	1 (6 %)	2 (7 %)	
	⇒ mucosal prolapse (revised)	1 (8 %)	3 (18 %)	4 (17 %)	

Table 2
CHD diagnosis sub-classification (n = 19).

Cardiac classification	Total
Non-duct-dependent CHD requiring surgery in the early neonatal period or infancy.	6 (32 %)
Left-to-right shunts undergoing surgery in infancy	3 (16 %)
Duct-dependent systemic circulation requiring biventricular neonatal repair	3 (16 %)
Duct-dependent pulmonary circulation requiring biventricular staged-surgery	3 (16 %)
Complex CHD anomalies	2 (11 %)
Pulmonary hypertension related to prematurity	1 (5 %)
Neonatal critical CHD	1 (5 %)

(Table 1). Of these, 19 patients (30.6 %) were identified as cardiac (Table 2). 18 cardiac patients (95 %) underwent at least one cardiac surgical intervention during the study period.

The median gestational age at birth was 38 weeks in both groups (p = 0.269). Mean birth weight did not significantly differ (2659 g vs. 2886 g, p = 0.351). Males predominated in both cohorts, comprising 79 % of the cardiac group (15/19) and 70 % of the non-cardiac group (30/43), though this difference was not statistically significant (p = 0.65). VACTERL association (≥3 anomalies) was significantly more prevalent in cardiac patients (n = 15/19 vs. 15/43, p < 0.001). Primary anoplasty was more frequently performed in non-cardiac patients (n = 7 vs. 1, p = 0.06).

3.2. ARM classification

ARM subtypes, categorised using the Krickenbeck classification, were similarly distributed across groups. Amongst female patients (n = 17), perineal fistulae and rectovestibular fistulae accounted for all cases. These subtypes were equally represented across both cardiac and non-cardiac cohorts. Amongst male patients (n = 45), rectobulbar fistulae were the most common subtype, affecting 16 (35 %) patients (6 cardiac, 10 non-cardiac) closely followed by perineal fistulae affecting 14 (31 %) patients (4 cardiac and 10 non-cardiac). Further subtypes included imperforate anus without fistula and rectobladder neck fistulae as detailed in Table 1.

3.3. Surgical timelines

The median age at stoma formation was 2 days in the cardiac group and 1 day in the non-cardiac group (p = 0.205). However, cardiac patients underwent definitive anorectal reconstruction significantly later (median 396 vs. 175 days, p = <0.001) even after

adjusting for birthweight and gestation ($p<0.002$) and age at stoma closure was therefore similarly delayed (554 vs. 265 days, $p < 0.001$). Cardiac patients also experienced a delay in stoma closure following completion of reconstruction compared with their non-cardiac counterparts (121 vs. 75 days, $p = 0.048$).

3.4. Complications

One or more postoperative complications were observed in 9/19 cardiac patients (47 %) compared with 12/43 non-cardiac patients (27 %). A total of 29 complications occurred across all time points (14 in the cardiac group, 15 in the non-cardiac group). While the odds of a cardiac patient experiencing 1 or more complication was 2.33 times higher than a non-cardiac patient (OR 2.33, 95 % CI 0.76−7.13), this was not statistically significant ($p = 0.136$).

Cardiac status was however a statistically significant predictor of total complication count (Wald $\chi^2 = 4.049$, df = 1, $p = 0.044$), with Poisson regression models showing cardiac patients had 2.11 times the rate of complications compared to non-cardiac patients (Incidence Rate Ratio/IRR = 2.11, 95 % CI: 1.23−4.37). When CHD and time to definitive reconstruction were modelled together, cardiac diagnosis was no longer a significant predictor of complication count (IRR 0.84, 95 % CI 0.34−2.10, $p = 0.709$), whereas surgical delay retained independent significance (IRR 1.002 per day, 95 % CI 1.000−1.004, $p = 0.019$).

Binary logistic regression showed each additional day between stoma formation to closure increased the odds of experiencing at least 1 complication by 0.4 % (OR 1.004, 95 % CI 1.000−1.008, $p = 0.029$).

Complication types were distributed across all time periods with no specific complication type predominating across stages or groups. Between stoma formation and definitive reconstruction (Time Period 1), 9 patients experienced adverse events, with unplanned, emergency laparotomy being the most common ($n = 5$, 3 cardiac, 2 non-cardiac). Three of these complications were encountered within 30 days of stoma formation. The complications were parastomal hernia requiring revision of stoma ($n = 1$), resection of mucus fistula perforation and re-fashioning of stoma ($n = 1$) and mucus fistula retraction/sepsis post-stoma formation ($n = 1$). Two complications occurred later but prior to definitive reconstruction; small bowel obstruction ($n = 1$) and stoma prolapse ($n = 1$). Other stoma-related complications included infection or dehiscence ($n = 2$) and elective stoma revision for ischaemic retraction of the mucous fistula ($n = 2$).

From definitive reconstruction to stoma closure (Time period 2), 9 patients suffered complications. These included wound dehiscence ($n = 2$) and need for re-do anoplasty ($n = 6$) either due to a stricture, retraction or mucosal prolapse. Around the time of stoma closure (Time period 3), complications were rarer and occurred in 5 patients. There was one anastomotic leak in a non-cardiac patient and two wound infections/dehiscence seen both in cardiac patients. Late complications (Time period 4) occurred in 7 patients and included neoanus stenosis requiring re-intervention ($n = 2$, 1 cardiac and 1 non-cardiac) and rectal mucosal prolapse requiring revision ($n = 4$, 1 cardiac and 3 non-cardiac).

4. Discussion

This study demonstrates that infants with anorectal malformations (ARM) and moderate to severe congenital heart disease (CHD) undergo definitive reconstruction at a median of 221 days (7 months and 11 days) later than their non-cardiac counterparts. This delay persists even after accounting for gestational age and birth weight. The detrimental impact of a cardiac diagnosis on outcomes in other congenital anomalies such as oesophageal atresia [16] and biliary atresia [17] are well recognised. In this later group, restorative cardiac surgery has been recommended prior to proceeding to reconstructive surgery for biliary atresia. The frequency of individual complications in our cohort was not large and we were therefore unable to identify any one predominant type of complication. Stenosis of the anoplasty can be considered an ischaemic complication potentially exacerbated by a cardiac diagnosis. However, only two cardiac patients suffered from this complication. One developed a significant stenosis prior to stoma closure and one patient post-stoma closure.

4.1. Surgical timing in CHD patients

Urgent decompression via stoma formation occurred at similar timepoints across groups, suggesting initial surgical acuity was not eclipsed by cardiac status. However, subsequent delays in reconstruction can be largely explained by the need for CHD patients to undergo cardiac surgery prior to their definitive reconstruction rather than more complications being encountered during Timepoint 1 (between stoma formation and definitive reconstruction). 18/19 (95 %) of cardiac patients required cardiac surgery during infancy with the similar proportion (31 % vs 29 %) of complications occurring during Timepoint 1 in both groups (4/14 vs. 5/15). Only one cardiac patient did not undergo cardiac surgery. This patient had significant pulmonary hypertension related to prematurity requiring optimisation prior to undergoing reconstructive surgery. Nonetheless, after discussing our findings with the cardiology team, there may have been an opportunity to potentially reconstruct some of the cardiac patients earlier. The delays in these cases largely result from institutional caution, prolonged anaesthetic and cardiology work-up before attaining clearance to proceed with resultant multidisciplinary hesitancy in the absence of formal protocols [3,5,15]. This is consistent with findings from Pelizzo et al. (2023), who reported that co-morbidity status, rather than ARM complexity, strongly predicted surgical timing [8].

Cardiac patients also suffered a significant delay in Timepoint 2 (between reconstruction and stoma closure); undergoing stoma closure at a median of 46 additional days compared with their non-cardiac cohort. This was again not explained by the complication rate being higher in the cardiac group during this time period (4/14 vs. 5/15). However, we postulate that additional preparation with the anaesthetic team to ensure cardiac optimisation prior to their stoma closure may have contributed to this delay.

Our data suggests that institutional decision paracpardcodeine making may be disproportionately influenced by the presence of a CHD label rather than functional severity or haemodynamic compromise. Several studies support this, noting that even minor CHD increases perioperative caution and resource allocation [3,15]. Furthermore, time taken for cardiology evaluation and multidisciplinary (MDT) planning may prolong surgical timelines irrespective of physiological need. Previous studies have highlighted the influence of institutional expertise and MDT consensus in determining operative readiness in CHD patients, particularly where formal guidelines are lacking [13].

4.2. Complications and time-dependent risk

CHD patients in our cohort experienced significantly higher complication rates, aligning with broader paediatric data linking congenital heart disease to poor post-operative outcomes due to impaired perfusion, immunity, and nutrition [13,14,18]. Our study suggests that complication risk may increase incrementally with surgical delay, with each additional day from stoma formation to closure conferring a 0.4 % rise in patient complication risk.

Instinctively, this risk seems to be wholly related to CHD diagnosis alone. Yet, when modelled together, cardiac diagnosis was no longer a significant predictor of complication count (IRR 0.84, 95 % CI 0.34−2.10, p = 0.709), whereas surgical delay to reconstruction retained independent significance (IRR 1.002 per day, 95 % CI 1.000−1.004, p = 0.019). This cautiously suggests that the effect of CHD on postoperative morbidity may be mediated by prolonged surgical timelines, rather than the cardiac condition alone. This reframes surgical delay not as a neutral waiting period, but as a continuous hazard exposure, particularly for physiologically vulnerable children. It further underscores the importance of prompt cardiac diagnosis and perioperative assessments in ARM infants. In CHD-ARM patients, delay may be viewed as an exposure that accrues risk, not simply a reflection of complexity. Time-sensitive approaches to surgical planning, which are gaining traction in other neonatal contexts [7,8], may prove useful when balancing urgency and safety in ARM-CHD patients.

4.3. Psychosocial and developmental consequences

Beyond immediate morbidity, delayed closure and reconstruction can extend the psychosocial impact of ARM. Families managing prolonged stoma care report increased stress, reduced social integration, and delayed toilet training [11]. Delayed reconstruction in our context, though sometimes clinically justified, may likewise extend caregiver stress, and lower health related quality of life scores, if not appropriately counselled and supported [8].

4.4. Limitations

Despite these associations, we emphasise that our data cannot establish causality. Surgical delay may, in many cases, be protective, allowing cardiopulmonary optimisation or family preparedness [13,14,18]. We could not capture the exact reasons behind surgical deferral, and institutional protocols may not reflect broader practices. Moreover, the small sample size limits the robustness of our regression analyses, and our exploratory statistics are likely influenced by unmeasured confounders which may more meaningfully account for increased complication risk. Important variables whose effect we cannot quantify here include;

- Inotropic or ventilatory burden
- Growth velocity or nutritional markers
- Anaesthetic and surgical team experience with CHD
- ICU resource availability and bed pressure
- Family or clinician preference
- Timing of cardiac investigations or interventional scheduling

Our retrospective design and single centre setting further limit generalisability. These limitations mirror those seen in prior observational studies of CHD risk in non-cardiac surgery and emphasise the need for structured, prospective data [5,15].

5. Conclusion

Cardiac ARM patients in our cohort experienced a significant delay in undergoing reconstructive surgery compared with their non-cardiac counterparts, with significantly increased complication rates. These delays occurred despite similar gestational age and birthweight. While causality cannot be inferred from this observational data alone, this study challenges the common presumption that deferring non-cardiac surgery in CHD patients is inherently protective, suggesting instead that delay may carry risks worth further exploration.

5.1. Clinical implications and future directions

Although deferment of reconstructive surgery may be appropriate in selected ARM/CHD cases, our data suggest that delay should not be assumed to be neutral or protective. Instead, surgical timing decisions should be explicitly framed as risk/benefit trade-offs, informed by a child's functional status rather than cardiac diagnosis alone, and families counselled accordingly.

Future research should prioritise:

- Prospective multicentre studies using standardised definitions of CHD severity e.g. ACS-NSQIP to guide surgical timing
- Development of surgical readiness tools that incorporate functional status, nutritional markers, and psychosocial context
- Functional outcome assessment in cardiac ARM patients

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Conflicts of interest

No Conflicts of Interest.

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