

*Full Length Research Paper*

## Late repair of truncus arteriosus improves the symptoms in the majority of patients: A systematic review

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Surgery aims to increase longevity and improve symptomatology. Surgical correction of truncus arteriosus after five months of age does not appear to offer a survival advantage over the natural history; repair after 5 months of age can be considered to be 'late' in this regard. Although surgical correction after five months of age can be achieved with low operative mortality, it remains unclear whether symptomatology is improved. Pubmed and Google scholar were searched between January 1st 1966 and September 30th 2022. Search terms included: Truncus, arteriosus, correction, repair, New York Heart Association, symptoms, and functional. Papers that included patients who had truncus arteriosus repair after the age of 5 months were examined. Three hundred and sixty seven results were obtained; 44 full-text papers were reviewed, seven full-text papers representing 16 patients were included. Surgical repair improved symptoms in 14 of 16 ( $87.5 \pm 18.3$ ; 95% CI) patients who had surgery after 5 months of age. Surgical correction of truncus arteriosus after 5 months of age improves symptoms in the majority of operable patients.

**Key words:** Pediatric, truncus arteriosus, operative, outcomes.

### INTRODUCTION

Truncus arteriosus (TA) is a form of cyanotic congenital heart disease characterised by a ventricular septal defect (VSD) and a common arterial vessel that supplies the systemic and pulmonary circulations (Russell et al., 2011). The gold standard of care is surgical correction in the neonatal period (Naimo et al., 2021). Surgical correction involves closure of the VSD and insertion of a conduit between the right ventricle and the pulmonary

arteries. Contemporary neonatal operative mortality is about 7% (Mastropietro et al., 2019). The chance of irreversible pulmonary vascular disease (IPVD) increases as the unoperated patient ages; the presence of IPVD renders the patient inoperable (Zhang et al., 2012). The goal of surgery is to increase longevity and improve symptomatology. With respects to symptoms, the parents of younger patients may report that their child feeds

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**Table 1.** Search strategy.

Search number and search terms
1. Truncus, arteriosus, repair (GS)
2. Truncus, arteriosus, correction (GS)
3. Truncus, arteriosus, symptoms (PM)
4. Truncus, arteriosus, functional, class (PM)
5. Truncus, arteriosus, New, York, heart, association (PM)

GS = Goggle scholar, PM = Pubmed.

Source: Author

**Table 2.** Full-texts included.

Author	Number of patients	Patients age (years)	Type of conduit	Post-operative symptoms
Gouton (Gouton et al., 2018)	1	3	Valved	OM
Ruan (Ruan et al., 2016)	1	33	Valved	Not improved (NYHA II)
Curi-Cur i(Curi-Curi et al., 2010)	4	<8	Valved	Improved in 3; OM in 1
Stegmann (Stegmann et al., 1982)	3	<3.6	Valved	Improved in all 3 patients
Griep (Griep et al., 1977)	4	<9	Valved	Improved in 3 of 4 patients
Rogers (Rogers et al., 1971)	1	10	Valved	Improved
Wallace (Wallace et al., 1969)	2	<9	Valved	Improved in both patients

OM = Operative mortality.

Source: Author

poorly or sweats while feeding. Older patients may report that they have tachypnoea or dyspnoea; particularly on exertion. The New York Heart Association (NYHA) classification is frequently used to describe a patient's functional capacity in terms of how symptomatic they become as they go about their daily activities. A recent study demonstrated that the surgical correction of TA in operable patients over 5 months, does not improve survival compared to the natural history (Awori et al., 2021). In view of this finding, we consider TA repair after 5 months of age as late TA repair. Surgical repair of TA after 5 months of age can be achieved with low operative mortality (Zhang et al., 2012). However, it remains unclear whether symptomatology is improved after late TA repair (Awori et al., 2021). This review aims to determine if late TA repair improves symptoms.

## MATERIALS AND METHODS

### Search strategy and selection criteria

Using the PRISMA-P checklist (Moher et al., 2015), PUBMED (PM) and Google Scholar (GS) were searched from January 1st, 1966 to September 30th, 2022. Table 1 shows the search terms and strategy. GS searches used "all in title" option; PM searches used "title/abstract" option. All search terms were combined with "AND". Titles and abstracts were reviewed and full-text articles were obtained when the abstract indicated that the paper might contain a description of preoperative and postoperative symptomatology on

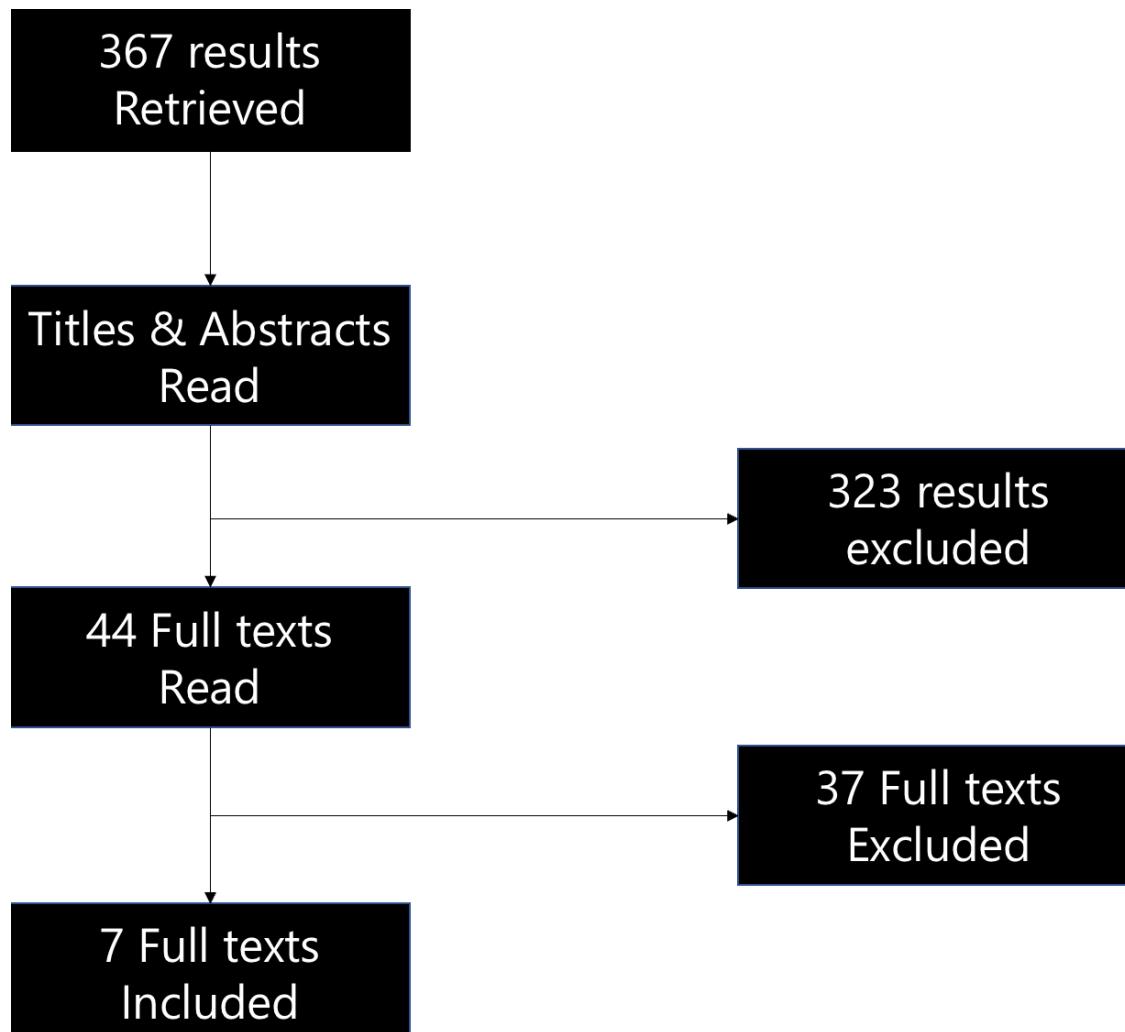
the same patient. Studies in a language other than English and non-human studies were excluded. Patients, who had 'truncal-valve' surgery or repair of concomitant cardiac lesions, with improvement of symptoms post-operatively, were excluded from our review.

## RESULTS

As shown in Figure 1, the search yielded 367 results; this was narrowed down further to 44 full-texts. Seven texts, representing 16 patients, describing pre- and post-operative symptoms were included in this review. Surgical correction improved symptoms in 14 out of 16 (85.7 +/- 18.3; 95% CI). There were two operative mortalities (12.5% +/- 16.2; 95% CI). Further details of these studies are shown in Table 2.

## DISCUSSION

The aim of surgical repair of TA is to increase longevity or to improve symptoms; preferably both. Earlier work by Awori et al. (2021), suggested that surgical repair of TA after 5 months of age may not offer a survival advantage over the natural history (Samánek, 1992). The question remained as to whether repair of TA after 5 months of age (late repair) would improve symptoms. After examining the existing literature, we found that the



**Figure 1.** Search flow diagram.  
Source: Author

surgical correction of TA in patients who presented later than 5 months of age, resulted in an improvement of symptoms in 14 out of 16 patients (87.5%). When the 95% confidence interval is taken into consideration however, there is a chance that the actual figure could be as low as 67.4%. Two out of 16 patients (12.5%) died in the early post-operative period. Although we examined a small group of patients, this mortality is similar to that reported by Naimo et al. (11.9%) for a larger sample of potentially similar patients (up to 8 years of age at the time of repair) (Naimo et al., 2021). Zhang et al. (2012) had no operative mortality in 12 patients who underwent 'late' repair. Although it seems obvious that surgery should improve symptoms, we were surprised at the dearth of work that actually reports preoperative and postoperative symptoms in the same patient. As surgical correction of TA has progressively been performed in younger and younger patients and has simultaneously been achieved with lower and lower operative mortalities

(Naimo et al., 2021). This development in conjunction with the general assumption that surgery improves symptoms for all operable patients, might explain why surgery is offered to operable late presenting patients and may also explain why few current studies examine symptomatology. Although it might seem obvious to experienced practitioners that surgery improves symptoms, we thought that it was important to examine the literature and document what evidence actually exists in this regard. We choose to examine preoperative and postoperative symptomatology in the same patient as opposed to comparing symptoms in operated and unoperated cohorts so that we could minimise confounding and determine how surgery would influence symptomatology in a particular patient.

Patients with TA may have concomitant cardiac lesions (such as an atrial septal defect or a persistent ductus arteriosus) or they may have had previous surgery such as pulmonary artery banding (Stegmann et al., 1982). In

either case, the lesions or the procedures may exacerbate the preoperative symptoms. In addition, the 'truncal' valve may have moderate or severe regurgitation, which further exacerbates symptoms. Moderate and severe truncal valve incompetence are indications for truncal valve repair (Naimo et al, 2021). To determine whether surgical repair of 'late presenting' TA improves symptoms, it was necessary to exclude patients who had truncal valve surgery or who had repair of potentially hemodynamically significant cardiac lesions. This further reduced the number of patients available for analysis in a subset of patients that was already limited.

Naimo et al. (2021) reported that most patients who had TA repair were in NYHA class I or II at long term follow-up; only one patient was in class III. The same paper reported that 68.6% of patients required at least 1 re-operation and 62.4% underwent a right ventricular outflow tract (RVOT) re-operation. The median time to this RVOT re-operation was 4.3 years. It is thought that a 16 mm diameter RVOT conduit can serve an adult patient; this size of conduit can successfully be placed in a 10 kg infant (Griep et al., 1977). Smaller sized conduits are a risk factor for RVOT re-operation (Naimo et al., 2021).

The anatomical arrangement in TA provides an abnormally high volume and pressure load to the pulmonary artery system. This results in progressive damage to the pulmonary arterial tree characterised by a progressive increase in pulmonary vascular resistance (PVR). Over time, this increase in PVR becomes irreversible. Traditionally, most centres will not operate on a patient whose indexed pulmonary vascular resistance (PVRI), after the administration of an inhaled pulmonary vasodilator, is greater than 8 Wood Units per meter squared ( $8 \text{ WU.m}^2$ ) (Zhang et al., 2012). This 'cut-off' for surgical repair was informed by the results of a study on 87 patients by Neutze et al. (1989). Further delays in surgical repair may result in a patient becoming 'inoperable' with regards to pulmonary vascular resistance.

Careful consideration is still required to determine if late surgery can be offered. If the actual percentage of patients who have a post-repair improvement of symptoms is 67.4%, it implies that about 3 out of every 10 patients who have late repair will not have improvement in symptoms. About 1 out of every 10 patients who have late repair dies from surgery. Even when early repair is undertaken, a number of patients are in NYHA class II at long term follow-up. It is assumed that this applies to late repair as well. Taken together these facts suggest that each patient (or patients' parents/guardians) will be required to weigh the 'pros' and 'cons' of surgery before accepting to have a late repair. This trade-off may be summarized as follows:

- (1) Late surgery may not improve symptoms in about 1 out of every 3 patients who survive surgery.
- (2) One out of every 10 patients operated on will die from

surgery.

(3) Two out of every 3 patients who survive the surgery will require another surgery about 4 or 5 years after the first surgery.

(4) Further delays in surgery may make the patient unsuitable for surgery (inoperable).

This review has some limitations; our sample size is small resulting in a broad 95% confidence interval. Nonetheless, we can be relatively certain that late correction will improve symptoms in at least 7 out of 10 patients; probably more. Some of the studies included are over 30 years old. The basic surgical technique (conduit repair) has not changed significantly over the years. Similarly, the medications used to treat symptoms postoperatively are fundamentally similar. For these reasons, we think that the results from these older studies remain valid in the current era. Since the criteria for operability were set using the work by Neutze et al. (1989), a much larger study by Gan et al. (2014) has been published. This work used more rigorous techniques to determine new criteria for operability; a PVRI of 10.3 WU after inhaling 100% oxygen was proposed as the 'cut-off' for operability (Gan et al., 2014). This implies that some patients, who would have been considered in-operable using the older criteria, can now be offered correction.

In conclusion, available evidence suggests that repair of TA after 5 months of age results in symptomatic improvement in the majority of patients. Late surgery does not appear to increase the operative mortality. Delays in surgery may render the patients inoperable.

## CONFLICT OF INTERESTS

The authors have not declared any conflict of interests.

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