Outcomes of descending and thoracoabdominal aortic repair in connective tissue disorder patients

Magnus Jonsson, Linus Blohm, Alireza Daryapeyma, Anders Günther, Göran Lundberg, Lena Nilsson, Carl-Magnus Wahlgren, Anders Franco-Cereceda and Christian Olsson

Department of Vascular Surgery, Karolinska University Hospital, Stockholm, Sweden; Department of Molecular Medicine and Surgery, Karolinska Institutet, Stockholm, Sweden; Department of Cardiothoracic Anesthesia, Karolinska University Hospital, Stockholm, Sweden; Department of Cardiothoracic Surgery, Karolinska University Hospital, Sweden

ABSTRACT

Objectives: Open surgical repair (OSR) of descending and thoracoabdominal aortic aneurysms carries risks of mortality and major complications. Patients with connective tissue disorders (CTD) are younger and require safe, efficient treatment with long-term durability. This study provides current outcome data to help inform treatment decisions.

Methods: All OSRs of descending thoracic aortic aneurysm (DTAA) or thoracoabdominal aortic aneurysm (TAAA) from January 2011 to July 2021 were included in a retrospective cohort study. Primary outcome measures were early and follow-up mortality and reintervention. Secondary outcome measures were major complications. Kaplan-Meier methods were used to estimate reintervention-free survival.

Results: A total of 26 OSRs (7 DTAA, 19 TAAA) were performed in 23 patients: 20 (77%) Marfan and 6 (23%) Loey-Dietz syndrome; median age 43 years. Aortic dissection was present in 100% and 3/26 (12%) were urgent. Early mortality was 1/26 (3.8%). No patient suffered spinal cord ischemia, stroke, vocal cord paralysis, or re-exploration for bleeding. The transient respiratory failure occurred in 19% (5/26) and transient renal replacement therapy in 15% (4/26). Renal function normalized in all patients within 3 months. During follow-up (median 4.6, range 0–11 years) there were no deaths and only one re-intervention on a previously operated aortic segment, resulting in 92% reintervention-free survival at 5 years.

Conclusions: In dedicated units, open surgical DTAA and TAAA repair in patients with CTD can be performed with a very low risk of death, severe complications and, late re-intervention. For CTD patients with reasonable risk, OSR should remain the first line of treatment.

ARTICLE HISTORY
Accepted 10 September 2022
Revised 17 August 2022
Received 15 March 2022

KEYWORDS
Connective tissue disorder; thoracoabdominal aortic aneurysm; surgical repair; outcomes; reintervention

Introduction

Thoracic endovascular aortic repair (TEVAR), especially fenestrated and branched solutions is increasingly employed in the treatment of descending and thoracoabdominal aortic aneurysms regardless of pathogenesis [1,2] and portended an increased role also in the treatment of patients with Marfan syndrome [3]. In the absence of randomized controlled trials, cohort studies have shown comparable, and in some instances superior short-term outcomes for TEVAR as opposed to open surgical repair (OSR) [4]. The long-term outcomes of TEVAR and fenestrated-/branched endovascular aortic repair (F/BEVAR) remain less well outlined, but the substantial need for re-intervention and persisting risk of aneurysm sac dilation and complications have been reported [5,6]. Such concerns are underscored in younger patients with longer life expectancy, i.e. longer time at risk. Patients with connective tissue disorders (CTD), such as Marfan, Loey-Dietz and vascular Ehlers-Danlos syndrome, are younger at presentation with descending thoracic aortic aneurysm (DTAA) or thoracoabdominal aortic aneurysm (TAAA). Their CTD bears an inherent risk for progressive aortic dilatation despite primary successful endovascular repair and also for specific TEVAR and F/BEVAR-related complications, e.g. stent graft-induced new entry tears and retrograde aortic dissection, respectively [7,8].

Given the increased re-intervention burden and as yet unproven long-term durability, TEVAR outcomes in CTD patients would need to be equal or surpass those of OSR if TEVAR is considered outside an emergency or bail-out-setting as suggested by current guidelines [9]. This study provides updated and detailed data on outcomes including mid-term follow-up, after OSR in CTD patients with DTAA or TAAA. The aim is to help inform adequate treatment selection and to establish a contemporary benchmark for comparison.

Patients and methods

During the ten-year study period (January 1, 2011 to July 1, 2021), in a university hospital unit providing OSR as well as TEVAR and F/BEVAR, a total of 115 OSRs and 356 endovascular repairs (TEVAR, n = 199 and F/BEVAR n = 157)

CONTACT Christian Olsson christian.olsson@ki.se Department of Cardiothoracic Surgery, Karolinska University Hospital, Eugeniavägen 23 C12:27, SE17176, Stockholm, Sweden

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for DTAA or TAAA were performed. Of these, 26 OSRs (22.6%) were performed in 23 CTD patients, forming the study cohort. Individual treatment modality was the result of shared decision-making in a multi-disciplinary conference. No patients were excluded. No CTD patients underwent primary endovascular repair. Pre-, intra- and postoperative variables were retrospectively collected from medical records to form the study database. Vital status was recorded on February 14, 2022 and was 100% complete. The study was approved by the national board of research ethics (No 2008/1771-31, 2020/02503), waiving the need to obtain written informed consent.

**Variable definitions**

Marfan syndrome was diagnosed clinically, using revised Ghent criteria, and/or with genetic panel testing. Loeys-Dietz syndrome was confirmed using genetic panel testing. Study variables were defined and outcomes were reported in accordance with current guidelines and standards of reporting [9,10]. Preoperative characteristics included comorbidities: hypertension, diabetes or chronic obstructive pulmonary disease, smoking status, preoperative creatinine level (µmol/L) and chronic renal failure, left ventricular ejection fraction, previous open heart surgery (with or without elephant trunk), the pathogenesis of aortic dilation (aneurysm vs chronic dissection), the extent of aortic dilation (DTAA vs TAAA type I-V), maximal aortic diameter (mm), and previous TEVAR or OSR in the same aortic segment (i.e. redo procedure). Intraoperative variables included perfusion (left heart bypass (LHB) vs full cardiopulmonary bypass (CPB) vs hypothermic circulatory arrest (HCA)), duration of extracorporeal circulation, use of cerebrospinal fluid drainage, reimplantation of visceral (coeliac trunk and superior mesenteric artery) and renal arteries, number of reimplanted intercostal arteries, and use of aorto-bi-iliac graft. Postoperative variables included in-hospital death, new stroke, new spinal cord injury, new renal failure requiring continuous renal replacement therapy, respiratory failure requiring prolonged (>72 h) intubation, reintubation, or tracheostomy, new vocal cord paralysis, and, re-exploration for bleeding. During follow-up, death, reinterventions on previously treated aortic segments, and recovery of creatinine levels (within 3 months of operation) were recorded and patients underwent repeat CT (occasionally MRI) of the entire aorta.

**Perioperative procedures**

Patients were managed by the same team of surgeons, anesthesiologists, and perfusionists. Intraoperatively, they were placed in a right decubital position, had a double-lumen endotracheal tube to allow one-lung ventilation, and were routinely provided with a cerebrospinal fluid drain connected to an automated peristaltic pump (LiquoGuard™, Möller Medical, Fulda, Germany) for monitoring and adjustment of cerebrospinal fluid pressure. Near-infrared spectroscopy (INVOS™ 5100, Medtronic, Minneapolis, Mn, USA) was used for neuromonitoring, with sensors placed bilaterally on the forehead and paraspinally approximately at the level of the 12th thoracic vertebra. All patients had, in addition to central venous lines, a central dialysis catheter placed for rapid volume transfusion and for postoperative continuous renal replacement therapy if needed.

All operations were performed through a left posterolateral thoracotomy. In TAAA, the incision was continued caudally in a paramedian plane, with the division of the costal arch and circumferential take-down of the diaphragm to aid exposure of the subdiaphragmatic aorta retroperitoneally with a self-retaining retractor.

The left femoral artery was always employed for arterial return, established through an 8-mm dacron graft to maintain lower extremity perfusion. For LHB, a left-sided pulmonary vein was cannulated with a 24 F angled cannula whereas, for CPB or HCA, the left femoral vein was cannulated with a 25 F multistage bicaval cannula to the right atrium. For LHB and CPB passive hypothermia was allowed, for HCA active cooling to 24°C core temperature was used, followed by HCA and insertion of perfusion cannulae in the cervical vessels for selective antegrade cerebral perfusion for the duration of the proximal anastomosis. Thereafter, an additional arterial cannula was connected to the graft side arm and reperfusion of the upper body begun. Full-dose systemic heparinization was always used and iterated to maintain activated clotting time >480 s.

Sequential aortic clamping from proximal to distal was employed, isolating one aortic segment each for the proximal anastomosis, reimplantation of intercostal arteries, reimplantation of visceral and renal arteries and for the distal anastomosis-es as necessary. The inferior mesenteric artery was not routinely reimplemented and accessory renal arteries were reimplemented when judged important and technically feasible. Visceral arteries were perfused with normothermic blood until reimplemented and reperfused. Renal arteries were perfused with hypothermic blood until reimplemented and reperfused. The choice of reimplantation technique (patch vs branched graft) was based on patient characteristics (age) as well as anatomical considerations (distance between Ostia) and intraoperative circumstances (aortic wall characteristics, time consumption, bleeding status) as exemplified in Figure 1.

Perioperative management included maintenance of MAP above 70 mm Hg for up to 72h postoperatively; maintenance of cerebrospinal fluid pressure below 15 mm Hg until the clinical assessment of neurological function was possible; liberal indication for dialysis to manage fluid overload and, early extubation and non-invasive ventilation as necessary.

**Statistics**

Data are presented as numbers with percentages and medians with ranges (min-max). The paucity of endpoint events precluded meaningful statistical analysis of variables associated with outcomes. Changes in creatinine levels were analysed with the Wilcoxon matched-pairs sign rank test. Survival and reintervention-free survival were estimated using Kaplan-Meier methods. Statistical analysis was performed using Stata v16 (Stata Corp, College Station, Tx, USA).
Results

During the study period, 23 patients underwent a total of 26 operations; 7 (27%) DTAA and 19 (73%) TAAA procedures, of which the majority (12/19) were extent II TAAA (Table 1). For the majority of operations (20/26) Marfan syndrome was the underlying CTD. No patient had vascular Ehlers-Danlos syndrome. Overall, findings in Marfan and Loeys-Dietz syndrome were similar (Tables 1–3). All patients had aortic dissection as underlying pathogenesis, approximately equally distributed between type A and type B as index diagnosis. Three operations were urgent; one re-dissection with intrathoracic aortic rupture, two de novo type B-dissections (one distal to a previous dacron graft in the descending aorta, one retrograde after open infrarenal aneurysm repair). The prevalence of substantial comorbidity was low, whereas, in almost all instances (24/26), previous open-heart surgery and/or TEVAR had been performed (Figure 2).

Aortic branch artery management and outcomes

Overall, 128 branch arteries (36 renal, 19 coeliac trunk, 19 superior mesenteric arteries, 1 inferior mesenteric artery, 53 intercostal or lumbar artery) were revascularized, i.e. reimplanted to the main graft either as part of a patch (8/19, 42%) or anastomosed individually or through a side-branch (11/19, 58%). Renal arteries were revascularized in 18/19 TAAA repairs, in one a beveled distal anastomosis was used. For renal arteries, coeliac trunk, and superior and inferior mesenteric arteries, 100% of targets were revascularized and the primary success rate was 74/75 (99%); one left renal artery side-branch anastomosis was revised. Intercostal arteries were reimplanted in 3/7 DTAA (all targets) and 14/19 TAAA (14/17 targets, 82%) repairs and in one case an already occluded lumbar artery was reopened and reimplanted to counteract intraoperative signs of spinal cord ischemia. All patients had a radiological follow-up with CT and/or MRI (range, 1–11 studies). One side-arm branch to an intercostal artery pair was occluded without clinical consequences, all other branches remained patent.

Primary endpoint (mortality)

Overall in-hospital mortality was 1/26 (3.8%); zero for non-urgent procedures. One patient with Marfan syndrome underwent open surgical repair of an infrarenal aortic
aneurysm approximately one month earlier; a procedure eliciting a retrograde dissection up to the level of the left subclavian artery, with rapid expansion, pleural effusion and renal failure but also deferred due to septicemia. He underwent sub-acute TAAA extent II repair under HCA, with ongoing hemodialysis preoperatively. Postoperatively, severe vasopoplegia developed, and extracorporeal membrane oxygenation was instituted, but vasopoplegia persisted and evidence of advanced brain injury mandated therapy withdrawal with ensuing death on a postoperative day 3. There were no other deaths, early or during follow-up (median 4.6 years, range 0–11 years, total 134 patient-years). For comparison, in-hospital mortality in the overall 10-year experience (n = 115) was 8/89 (9.0%) in non-CTD patients, with a similar distribution for DTAA and TAAA, respectively (Figure 3).

**Table 1.** Demographic and preoperative characteristics in 26 open surgical DTAA and TAAA repairs in patients with connective tissue disorders.

| Characteristic                  | All (n = 26) | Marfan (n = 20) | Loeys-Dietz (n = 6) |
|--------------------------------|--------------|-----------------|---------------------|
| Male                           | 17 (65)      | 13 (65)         | 4 (67)              |
| Age, years                     | 43 (15–63)   | 49 (38–63)      | 35 (15–52)          |
| Height, cm                     | 191 (166–204) | 191 (166–204)  | 176 (170–200)       |
| Weight, kg                     | 82 (40–124)  | 81 (64–124)     | 84 (40–95)          |
| BMI, kg/m²                     | 23 (14–34)   | 23 (17–34)      | 22 (14–32)          |
| Hypertension                   | 13 (50)      | 13 (65)         | 0                   |
| Smoking                        | 0            | 0               | 0                   |
| COPD                           | 0            | 0               | 0                   |
| Diabetes                       | 1 (4)        | 0               | 1 (17)              |
| LVEF, %                        | 55 (40–60)   | 55 (40–60)      | 55 (50–55)          |
| Renal failure                  | 1 (4)        | 1 (5)           | 0                   |
| S-creatinine, µmol/L           | 78 (40–233)  | 75 (43–233)     | 88 (40–126)         |
| Chronic dissection             | 26 (100)     | 20 (100)        | 6 (100)             |
| Type A                         | 15 (58)      | 11 (55)         | 4 (67)              |
| Type B                         | 11 (42)      | 9 (45)          | 2 (33)              |
| DTAA                           | 7 (27)       | 6 (30)          | 1 (17)              |
| TAAA                           | 19 (73)      | 14 (70)         | 5 (83)              |
| Extent I                       | 0            | 0               | 0                   |
| Extent II                      | 12 (46)      | 8 (40)          | 4 (67)              |
| Extent III                     | 5 (19)       | 4 (20)          | 1 (17)              |
| Extent IV                      | 1 (4)        | 1 (5)           | 0                   |
| Extent V                       | 1 (4)        | 1 (5)           | 0                   |
| Max aortic diameter, mm        | 55 (50–70)   | 55 (50–70)      | 54 (50–56)          |
| Previous open heart surgery    | 24 (92)      | 19 (95)         | 5 (83)              |
| Elephant trunk                 | 7 (27)       | 5 (25)          | 2 (33)              |
| TEVAR                          | 2 (8)        | 2 (10)          | 0                   |
| Redo procedure                 | 5 (19)       | 4 (20)          | 1 (17)              |
| Urgent/subacute                | 3 (12)       | 3 (15)          | 0                   |

Data are presented as n (%) or median (range).

**Table 2.** Intraoperative procedures and characteristics in 26 open surgical DTAA and TAAA repairs in patients with connective tissue disorders.

| Procedure                           | All (n = 26) | Marfan (n = 20) | Loeys-Dietz (n = 6) |
|-------------------------------------|--------------|-----------------|---------------------|
| Perfusion strategy                  |              |                 |                     |
| Cardiopulmonary bypass              | 2 (8)        | 1 (5)           | 1 (17)              |
| Hypothermic circulatory arrest      | 6 (23)       | 5 (25)          | 1 (17)              |
| Left heart bypass                   | 18 (69)      | 14 (70)         | 4 (67)              |
| Cerebrospinal fluid drain           | 22 (85)      | 16 (80)         | 6 (100)             |
| Extracorporeal circulation, min     | 187 (69–343) | 179 (69–343)    | 195 (184–241)       |
| Visceral artery reimplantation      | 19 (73)      | 14 (70)         | 5 (83)              |
| Renal artery reimplantation         | 18 (69)      | 13 (65)         | 5 (83)              |
| Number of intercostal arteries reimplanted | 2 (0–6)   | 2 (0–4)         | 4 (2–6)             |
| Aorto-bi-iliac graft               | 6 (23)       | 5 (25)          | 1 (17)              |

Data are presented as n (%) or median (range).

**Secondary endpoints (major complications)**

No patient had spinal cord ischemia, stroke, permanent dialysis, tracheostomy, vocal cord paralysis or re-exploration for bleeding (Table 3). Notably, none of the major complications could be definitively determined in the patient expiring on the third postoperative day. Renal function deteriorated temporarily with a significant median 1.56-fold increase of serum-creatinine but recovered fully (normalized serum-creatinine) in all surviving patients within 3 months postoperatively (Figure 4). Hospitalization was relatively short: median 9 days, range 6–20 days including a stay in intensive care for a median of 4 days, range 2–20 days. In non-CTD patients, spinal cord ischemia developed in 3/89 (3.4%), again with similar distribution between DTAA and TAAA (Figure 3). Renal failure requiring new-onset continuous renal replacement therapy occurred in 4/19 (21%) TAAA repairs only, whereas transient respiratory failure occurred in 4/19 (21%) TAAA and 1/7 (14%) DTAA repairs, respectively.

**Follow-up reinterventions**

One late re-intervention of a previously operated aortic segment occurred; a visceral artery patch anastomotic pseudoaneurysm developed 1.5 years postoperatively in a 56-year-old woman with Marfan syndrome. She was successfully treated with a branched endo-prosthesis. In all other patients, CT/MR surveillance did not show signs of evolving complications in treated aortic segments or branches. Thus, survival free of unplanned re-intervention was 92% at 5 years (Figure 5) and the total event (death or reintervention) rate was 1.4 per 100

**Table 3.** Postoperative outcomes after 26 open surgical DTAA and TAAA repairs in patients with connective tissue disorders.

| Procedure                           | All (n = 26) | Marfan (n = 20) | Loeys-Dietz (n = 6) |
|-------------------------------------|--------------|-----------------|---------------------|
| In-hospital mortality               | 1 (4)        | 1 (5)           | 0                   |
| Follow-up mortality                 | 0            | 0               | 0                   |
| Spinal cord ischemia                | 0            | 0               | 0                   |
| Stroke                              | 0            | 0               | 0                   |
| Temporary dialysis                  | 4 (15)       | 2 (10)          | 2 (33)              |
| Permanent dialysis                  | 0            | 0               | 0                   |
| Respiratory failure                 | 5 (19)       | 3 (15)          | 2 (33)              |
| Tracheostomy                        | 0            | 0               | 0                   |
| Re-exploration                      | 0            | 0               | 0                   |
| Vocal cord paralysis                | 0            | 0               | 0                   |
| Late reintervention                 | 1 (4)        | 1 (5)           | 0                   |

Data are presented as n (%).
patient years. Three other patients underwent planned aortic operations on previously non-operated aortic segments, as part of planned staged procedures.

Discussion

This contemporary cohort of 26 DTAA and TAAA open repairs in patients with CTD presented excellent outcomes, with only one early death (none in the elective setting), no additional deaths during follow-up and, one single unplanned reintervention during a median of nearly five years follow-up. Severe complications, such as spinal cord ischemia, stroke, re-exploration for bleeding and permanent renal failure, were absent altogether. Visceral arterial patch reimplantation was employed in 42% of TAAA procedures and resulted in one late reintervention. While it should be generally discouraged due to the risk of aneurysm formation, failures were not abundant and its use is based on the weighing of several intraoperative factors. Nevertheless, we too have abandoned this practice in favour of separate reimplantation.

Previously reported CTD patient cohorts undergoing OSR for DTAA and TAAA have shown similarly satisfying early and mid-term outcomes [11–15] (Appendix Table 1). Death and severe complications are with few exceptions reported with zero or single-digit prevalence. Additionally, a low prevalence of death and reintervention (16% at six years postoperatively) as well as virtually normal quality-of-life status has been demonstrated with OSR [16]. While these results were achieved in dedicated high-volume centers, the findings of the present study emanated from a non-high-volume center [17], suggesting that the broader generalizability of the findings is reasonable, but as yet confined to units with expertise in OSR for DTAA and TAAA.

TEVAR, including F/BEVAR, in CTD patients, has been employed. While the technical success rate has been very high (96–100%) [18] and early outcomes (mortality, spinal cord injury, major complications) favorable in the 0–15% range [6, 18], mid-term complications have also been abundant. Overall primary treatment failure during shorter (less than two years) follow-up approaches is 45% and include device migration, endoleak of variable (sometimes multiple) origin, branch occlusion or dysfunction, re-resection (including stent graft-induced new entry and retrograde dissection), false-lumen expansion and aortic dilation with associated lethal complications such as rupture and aorto-bronchial fistulae [6,18–20]. Nevertheless, endovascular repair techniques continue to evolve, and more selective approaches may contribute to improved outcomes [21–23]. Importantly, while secondary OSR after simple TEVAR can be performed with acceptable outcomes (except for graft infection and fistulation) [24,25],
procedures mandating full or partial extraction of branched stent grafts will pose a substantial risk for harm. Lastly, analyzing the cost-effectiveness of fenestrated and branched TEVAR in complex TAAA has proven difficult, due to the lack of high-quality data [26]. Several studies suggest the cost of multiple endovascular devices is the primary, unavoidable driver for high TEVAR costs [27,28]. This effect is more pronounced in CTD, characterized by younger patients with a substantial need for follow-up and reintervention, but also with generalized arterial disease often ending up with (sub)total replacement of the entire aorta, as perceived from Figure 2. Conversely, in OSR, complications and prolonged hospitalization drive costs. In the present study, renal and respiratory failure were relatively uncommon and rapidly resolving. Other major complications approached zero prevalence. This was reflected by a short median length of hospital stay of 9 days, and suggests that, for CTD patients, OSR in reasonable candidates will also retain superior cost-effectiveness compared to endovascular repair.

**Study limitations**

The shortcomings of small retrospective cohort studies are applicable and interpretations should be cautious. The present study is descriptive. CTD patients constituted 21% of all DTAA and TAAA OSR during the study period. No CTD patient underwent primary TEVAR or F/BEVAR during the same period, hence the lack of comparison. OSR in non-CTD patients is characterized, among other things, by older age, variable pathogenesis, increased comorbidity and lower proportions of extent II TAAAs and previous aortic interventions and were therefore not considered a relevant group for comparison of outcomes. DTAA and TAAA were grouped, the latter constituting around 75%. Given the considerably larger extent of OSR for TAAA, findings should not be interpreted as applying to TAAAs alone.

To summarize, early outcomes can be equally good with OSR as with TEVAR in CTD; death and devastating complications are infrequent with both approaches. True long-term data are generally scarce, but at medium-term follow-up, the need for re-intervention on previously treated segments of the aorta appears uncommon after OSR but much more frequent after TEVAR; in turn, adding risks as well as a cost without apparent advantage. Therefore, outside well-designed and controlled randomized trials, the current guideline recommendations seem adequate: OSR is preferred for reasonable surgical candidates with CTD and TEVAR is
reserved for emergency and bail-out procedures, as a bridge to definitive treatment, and as secondary interventions to treat (the less frequent) late surgical failures [9]. Ideally, CTD patients with DTAA or TAAA should be managed by multi-professional aortic teams experienced in clinical decision-making as well as in performing any treatment modality assigned to benefit the individual patient most.

Disclosure statement

No potential conflict of interest was reported by the author(s).

Funding

The study was supported by a personal donation (to AFC and CO) from Mr Fredrik Lundberg.

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Appendix

Appendix Table 1. Summary of outcomes in recently published patient cohorts of open surgical repair of descending and thoracoabdominal aneurysms in patients with connective tissue disorders.

| Author          | Study period | n    | 30d mort | SCI | CRRT  | Stroke | Bleeding | Vocal cord | LOS |
|-----------------|--------------|------|----------|-----|-------|--------|----------|------------|-----|
| Mommertz et al. | 2000–2006    | 22   | 0        | 0   | 0     | 0      | 1 (4.5)  | n/a        | 24  |
| Coselli et al.  | 2004–2014    | 127  | 5 (3.9)  | 5 (3.9) | 6 (4.7) | 1 (0.8) | 10 (7.9) | 43 (34)    | 12  |
| Hicks et al.    | 2006–2015    | 29   | 3 (10)   | 1 (3.4) | 2 (6.9) | 2 (6.9) | 0        | n/a        | n/a |
| Keschenau et al.| 2000–2016    | 72   | 9 (12)   | 4 (5.5) | 7 (9.7) | 2 (2.8) | 20 (28)  | 6 (8.3)    | 22  |
| Frankel et al.  | 2006–2014    | 155  | 2 (1.3)  | 6 (3.9) | 8 (5.2) | 3 (1.9) | 15 (9.7) | 33 (21)    | 10  |
| Present study   | 2010–2021    | 26   | 1 (3.8)  | 0   | 4 (15) | 0      | 0        | 0          | 9   |

Numbers with percentages. Numbers in brackets indicate article reference numbers.

Abbreviations. 30d mort, 30-day mortality; CRRT, continuous renal replacement therapy; LOS, length of stay in hospital; SCI, spinal cord injury.