

A review of the peri-operative management of pediatric patients with connective tissue disorders undergoing cardiac surgery

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INTRODUCTION

Children with Marfan and Loeys-Dietz Syndromes (LDS) are at high risk for cardiovascular disease and frequently undergo cardiothoracic surgery in childhood and adolescence. There are a number of comorbidities described in both patient populations that may complicate their perioperative management. At present there are limited studies describing intraoperative management of pediatric patients with connective tissue diseases and none detailing those undergoing cardiac surgery. We aim to characterize this population and describe potential concerns regarding their intraoperative management.

METHODS

Using our institutional STS database, we identified a population 35 LDS and 42 Marfan patients who underwent cardiopulmonary bypass from 2011-2017. We completed a retrospective chart review, including the anesthetic record, to obtain demographic, medical and intraoperative data for each patient. We completed a simple descriptive analysis based on the data obtained.

RESULTS

Using retrospective chart review, we collected demographic and intraoperative data as described in tables 1 and 2, respectively.

Table 1

	Loeys-Dietz Syndrome	Marfan Syndrome
Male, n (%)	18 (51)	28 (67)
Age at time of surgery (IQR)	13 (9-17)	16 (11.75-18)
BMI (IQR)	16.8 (14.3-21.4)	18.65 (15.4-22.3)
NYHA Heart Classification, n (%)		
I	28 (82)	33 (80)
II	4 (12)	7
III	2 (6)	1 (3)
Ventricular dysfunction, n (%)		
Systolic	3 (9)	5 (12)
Diastolic	2 (6)	1 (2)
Number of previous cardiac surgeries, n (%)		
0	19 (54)	35 (83)
1	9 (26)	6 (14)
2	4 (11)	1 (3)
4 or greater	3 (9)	0
Preop ACE inhibitor, n (%)	34 (100)	36 (86)
Preop beta blocker, n (%)	14 (41)	28 (67)
Aortic root Z score (IQR)	4.27 (2.25-8.82)	6.58 (4.85-9.08)
Any valvular disease, n (%)	31 (89)	33 (80)
Bicuspid aortic valve	5 (14)	3 (7)
Aortic valve disease	15 (43)	13 (32)
Mitral valve prolapse	7 (20)	25 (61)
Mitral valve disease	9 (26)	23 (56)
Tricuspid valve disease	11 (31)	17 (41)
Pectus present, n (%)	18 (53)	16 (38)
Pulmonary comorbidities, n (%)	3 (9)	7 (17)
Medications, n (%)		
GI (Reflux/Constipation)	10 (32)	4 (10)
Pulmonary (asthma/allergy)	20 (64)	5 (12)
Pain	5 (16)	4 (10)
Obstructive sleep apnea, n (%)	4 (12)	1 (2)

Table 2

	Loeys-Dietz Syndrome	Marfan Syndrome
Type of surgery, n (%)	4 (12%)	
VSSR	22(63)	30 (71)
MV repair/replacement	2(6)	12 (28.5)
AV repair/replacement	4(11)	3(7%)
Other	9 (25)	2 (5)
Difficult airway, n (%)	7 (20)	0 (0)
CPB time (IQR)	141 (119-170)	143 (127.75-173)
Cross clamp time (IQR)	91 (68-119)	100.5 (80.75-114.75)
Intraoperative vasopressor use (%)		
Any Pressor	64	81
Epinephrine	52	75
Vasopressin	33	36
Milrinone	18	11
Blood products given, median # units (IQR)		
pRBC	1 (0-4)	0 (0-1)
FFP	0 (0-2)	0 (0-1)
Platelet	0 (0-1)	0 (0-0)
Cryo	0 (0-0)	0(0-0)
Extubated in the OR (%)		
Total	26	23
Since 2016	50	50
Peak lactate (IQR)	2.9 (2.35-5.1)	2.7 (2.1-3.675)
Post-op hemoglobin (IQR)	10.7 (9.975-11.925)	11.3 (9.475-12.5)
LOS (IQR)	7 (5-9.25)	6.5 (6-9)

DISCUSSION

These data reflect many years of experience at our institution and fill a gap in the existing literature. There were a number of findings for both patient populations that could affect anesthetic management. The most pertinent for LDS patients include the high percentage with a difficult airway or pectus; on ACE inhibitors, beta-blockers, GI, pulmonary and pain medications; recent extubation rate; and low BMI. The most pertinent for the Marfan patients include the high percentage with a pectus, receiving chronic pain medications, ACE inhibitors, beta-blockers, and relatively low blood utilization. Future directions include analysis of how these characteristics affected these patients during the entire perioperative period including the ICU stay.

CONCLUSIONS

LDS and Marfan patients undergoing cardiac surgery have a number of concomitant medical concerns that may affect their anesthetic plan.

REFERENCES

1. Loeys BL et al. *Nat Genet.* 2005 Mar;37(3):275-81.
2. Castellano J et al. *Seminars in Thoracic and Cardiovascular Anesthesia.* 2014;18(3):260-271.