

Anaesthetic and obstetric outcomes in pregnant women with Marfan syndrome

Dr Stephen Thompson, Ms Tonicka Virdee, Dr Kailash Bhatia

Saint Mary's Hospital, Manchester University NHS Foundation Trust



Introduction

- Marfan syndrome (MFS) is a connective tissue disorder affecting one in every 5000 individuals.
- Pregnant women with MFS may have cardiac (e.g. aortopathy) and specific musculoskeletal features (e.g. scoliosis, dural ectasia) that may have significant anaesthetic and obstetric implications [1].
- The aim of this service evaluation was to determine the anaesthetic and obstetric outcomes in pregnant women with MFS.

Methods

- We retrospectively reviewed records of 11 pregnancies in nine pregnant women with genetically confirmed MFS that received multidisciplinary input from the cardio-obstetric team between 1 January 2015 to 31 December 2024 at our tertiary centre.

Results

- From 11 pregnancies reviewed, 10 live births were recorded. One woman had a miscarriage following an aortic dissection. Their demographics are reported in table 1.
- Five of the nine (55.6%) women had scoliosis with two reported to have a surgical correction. Six of the nine (66.7%) were taking beta blockers intrapartum. Dural ectasia was not noted in any patient.
- Of the 10 live births, six women (60%) had a vaginal birth - three were spontaneous with three requiring an instrumental delivery (Figure 1).
- Neuraxial anaesthesia for caesarean birth was administered to all four women but one required conversion to general anaesthesia (GA) due to inadequate block height (Figure 2).
- The mean dose of intrathecal bupivacaine for the caesarean birth was 17 (± 2.71) mg.
- The postpartum maternal outcomes are highlighted in table 2.
- The mean neonatal birthweight was 3110 (± 515) g with Apgar scores highlighted in Figure 3.

Maternal demographics	Mean ($\pm$ SD)
Age (years)	29.4 (± 6.96)
Height (cm)	176.4 (± 8.10)
Body Mass Index (kg/m ²)	24.9 (± 4.58)
Pre-pregnancy maternal aortic root diameter (mm)	37.4 (± 3.37)

Table 1: Maternal demographics (n=10)

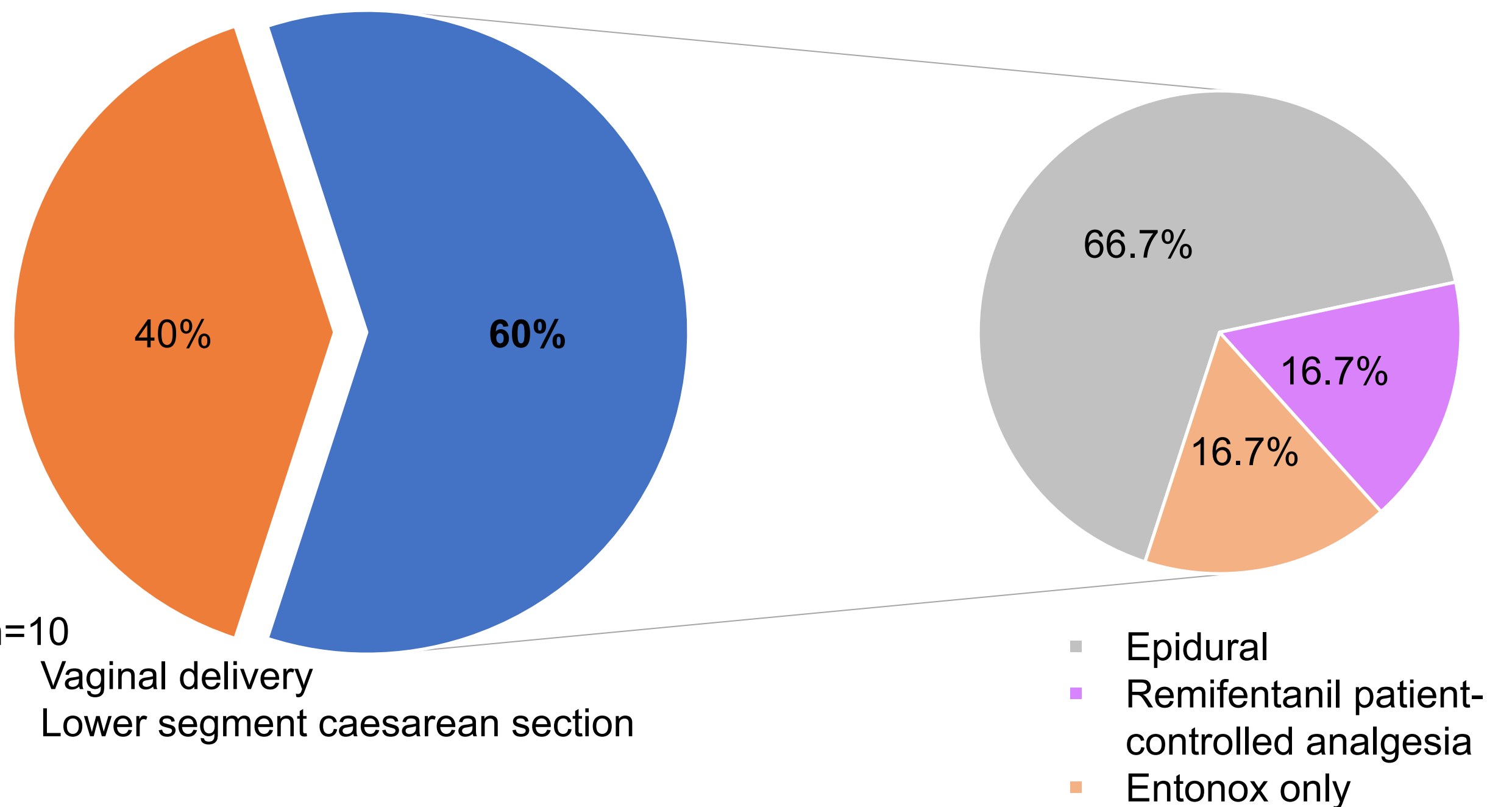


Figure 1: Mode of delivery and anaesthesia types in women who delivered vaginally and went into labour

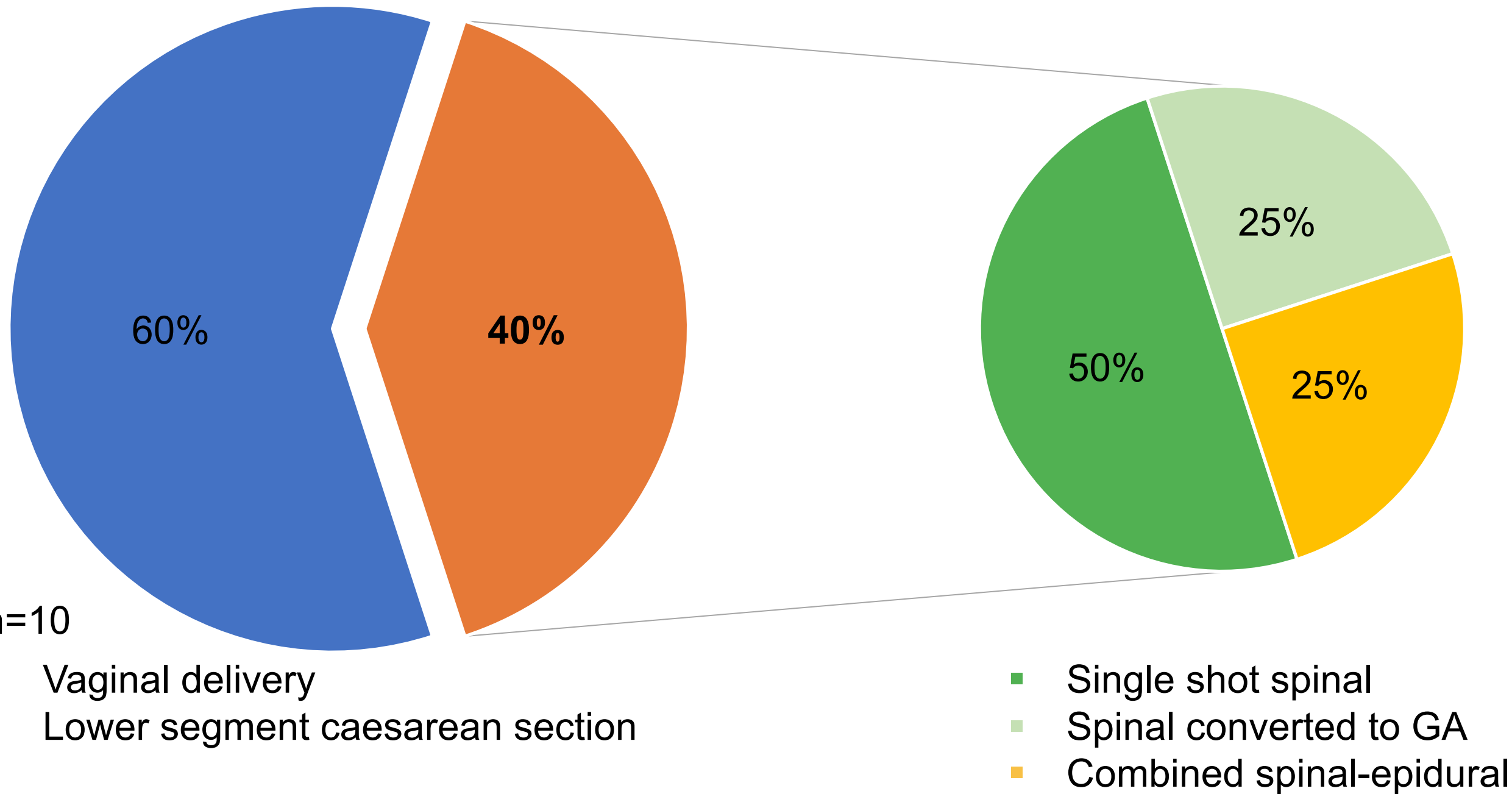


Figure 2: Mode of delivery and anaesthesia types for caesarean birth

Maternal outcomes	n (%)
Postpartum haemorrhage (>500ml)	3 (30%)
Mean length of hospital stay (>48 hours)	8 (80%)
High dependency unit admission	1 (10%)
Postpartum maternal aortic root diameter increase	2 (20%)

Table 2: Postpartum maternal outcomes (n=10)

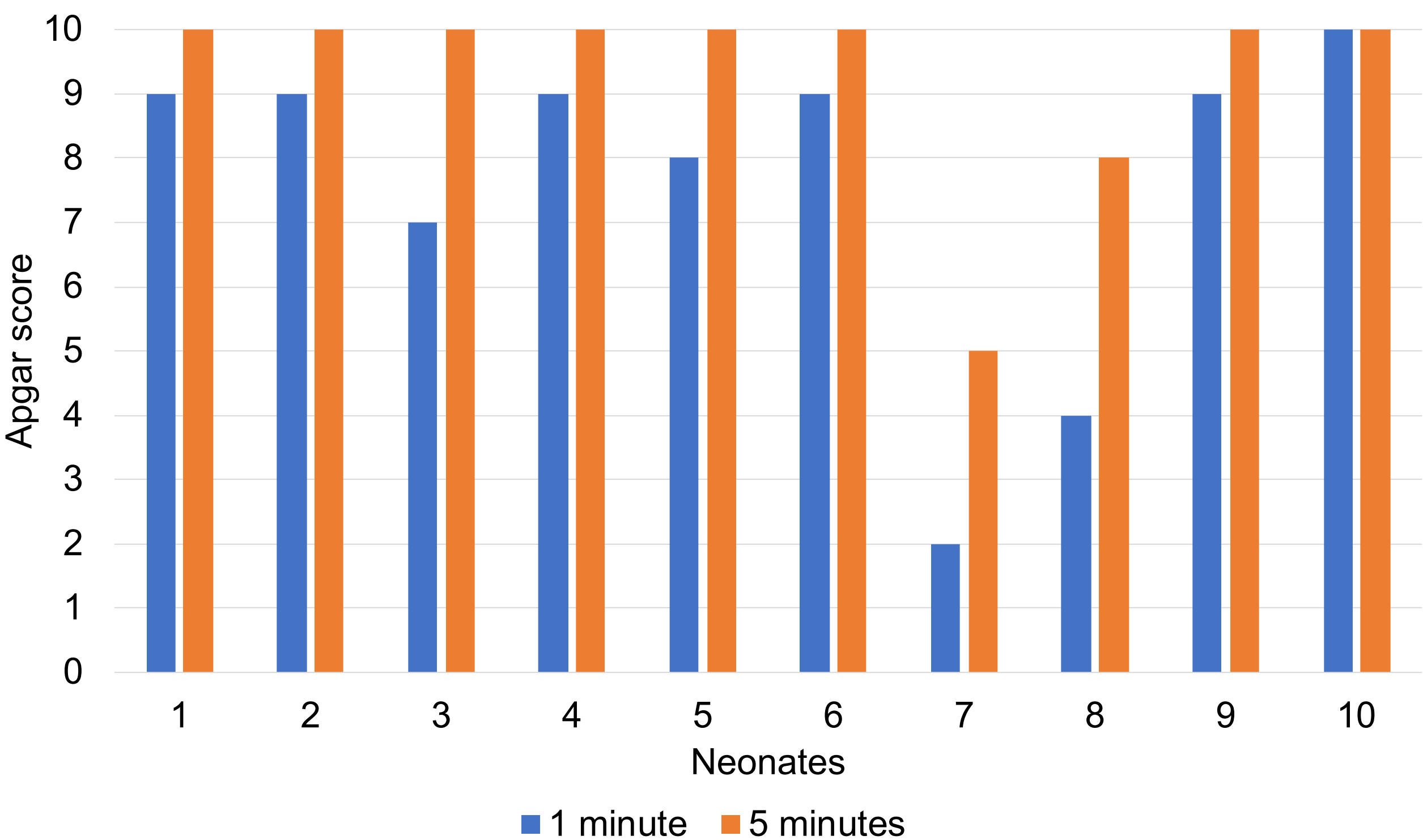


Figure 3: Apgar scores at 1 and 5 minutes for each live birth

Conclusion

- Our study highlights that although obstetric outcomes in MFS are favourable, the multidisciplinary cardio-obstetric team need to be prepared for rare cardiac emergencies including aortic dissection alongside more common emergencies such as postpartum haemorrhage.
- Higher than usual intrathecal local anaesthetic doses for caesarean birth are observed possibly due to scoliosis, women being relatively tall and inadequate neuraxial anaesthesia reported in literature on MFS [2].
- Pre-conception counselling should include information on the cardiac and obstetric risks but also highlight musculoskeletal issues that may lead to inadequate neuraxial analgesia and anaesthesia.

References

- [1] – Allyn J, Guglielminotti J, Omnes S, et al. Marfan's syndrome during pregnancy: anesthetic management of delivery in 16 consecutive patients. *Anaesthesia & Analgesia*. 2013; **116**: 392–8.
- [2] – Heck A, Clayton R, Robinson C. Dural ectasia in Marfan syndrome. *International Journal of Obstetric Anaesthesia*. 2014; **23**: 287-8.