

Anesthetic management in Marfan’s syndrome

¹Dr. Aishwarya Kini M, Junior Resident, A.J. Institute of Medical Sciences, Mangalore

²Dr. Vasanth Shetty, Senior Resident, A.J. institute of Medical Sciences, Mangalore

³Dr. Archana Srinivasan, Junior Resident, A.J. Institute of Medical Sciences, Mangalore

⁴Dr. Abhiram Shastry, Junior Resident, A.J. institute of Medical Sciences, Mangalore

Corresponding Author: Dr. Aishwarya Kini M, Junior Resident, A.J. Institute of Medical Sciences, Mangalore

How to citation this article: Dr. Aishwarya Kini M, Dr. Vasanth Shetty, Dr. Archana Srinivasan, Dr. Abhiram Shastry
“Anesthetic management in Marfan’s syndrome”, IJMACR- April - 2023, Volume – 6, Issue - 2, P. No. 566 – 568.

Open Access Article: © 2023, Dr. Aishwarya Kini M, et al. This is an open access journal and article distributed under the terms of the creative common’s attribution license (<http://creativecommons.org/licenses/by/4.0>). Which allows others to remix, tweak, and build upon the work non-commercially, as long as appropriate credit is given and the new creations are licensed under the identical terms.

Type of Publication: Case Report

Conflicts of Interest: Nil

Abstract

Background: Marfan syndrome is an autosomal dominant disease. It is seen in an incidence of 1 in 3,000 to 5,000 individuals. It is a multisystem disease which involves ocular (lens dislocation, myopia), cardiovascular (aortic root dilatation with aortic regurgitation, mitral valve prolapses with mitral regurgitation), and musculoskeletal abnormalities (long bone overgrowth, scoliosis, kyphosis, joint hypermobility), lung (pneumothorax), skin (striae), and central nervous system (dural ectasia). [1] General anesthesia in these patients poses a risk for hypertensive responses with increases risk for aortic dissection. They are also at risk for pneumothorax secondary to positive pressure ventilation. [1] Neuraxial anesthesia is a safer alternative but can however be complicated due to presence of skeletal deformities like kyphoscoliosis. [1] The challenge of administering neuraxial blockade can be overcome by usage of ultrasound to guide the

direction and location of block. The device is portable and relatively inexpensive and can be performed by an experienced expertise.

Case report

A 45-year-old male, weighing around 40 kgs, who was a known case of “MARFAN’S SYNDROME”, presented with inability to move his right lower limb due to slip & fall. He was diagnosed as right proximal femur fracture & was posted for “dynamic hip screw fixation”. On pre-anesthetic examination, patient had pallor, severe thoracolumbar kyphoscoliosis, deviation of trachea, pectus excavatum, arachnodactyly, B/L crepitations and decreased breath sounds over B/L basal areas, ESM murmur. On local examination of the lumbar spine, interspinous space was not felt X-ray of whole spine showed kyphoscoliosis of thoracolumbar spine with Cobb angle of 45°.

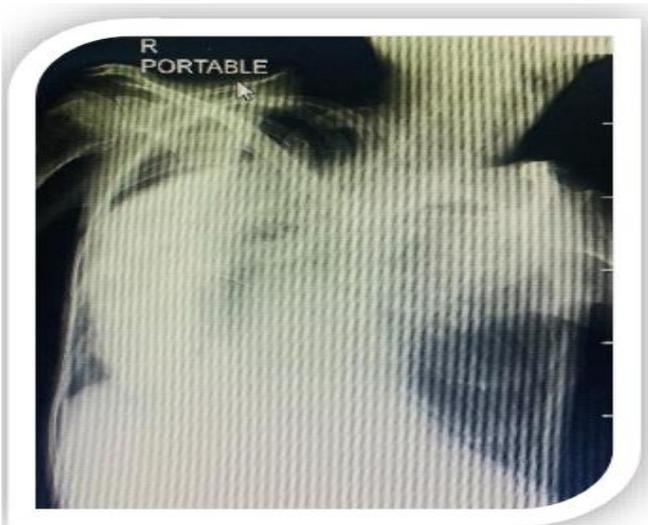


Figure 1

On airway examination: Mallampati grade 2 with adequate mouth opening with high arched palate was noted.

Pulmonary function test interpreted obstructive and restrictive lung disease with forced vital capacity and forced expiratory volume in 1s of 70% and 50% predicted values respectively.

ECHO shows: Mitral regurgitation & mitral valve prolapse, LVEF-60%. Pre-op investigation: Hb-7.8 gm/dl, Tc- 12,900, S.cr – 0.6 mg/dl & other parameters were within normal limits.

Patient was pre-operatively managed by transfusing 1-unit PRBC & started on nebulization with alpha agonist & incentive spirometry. Surgery was planned under spinal anesthesia.



Figure 2 and Figure 3

In the OR, multi-parameter monitors were connected & pre-loaded with Ringer lactate solution(15ml/kg).

To facilitate positioning for SAB, blind fascia Iliaca block was given-10cc of 0.25% Levobupivacaine.

In sitting position, under sterile aseptic conditions, parts painted & draped. Under USG guidance, the midline, L3-4 space and depth of curvature of spine was confirmed.

After two attempts, successfully lumbar puncture was performed at L3-L4 intervertebral disc space under USG guidance using 25G Quincke needle. After confirmation of free flow of CSF, 2CC(10mg) OF 0.5% heavy bupivacaine with fentanyl 20 mcg was injected. No vascular tap. Patient was made supine with mild right lateral tilt.

Hypotension, intraoperatively was managed by vasopressors. Intra-operative course was uneventful with blood loss of 300 ml. post-operatively, one PRBC was transfused. In post-operative period, spinal anaesthesia wore off in 2 hours.

Discussion:

Kyphoscoliosis is characterised by progressive deformity of spine. The level of derangements in cardiac and pulmonary function of kyphoscoliosis patient is related to the amount of cobb's angle in thoracolumbar X-ray. This causes difficulty for anaesthetic practitioners for administration of both general and spinal anaesthesia. Neuraxial anaesthesia in this type of patient is technically challenging. The causes of failed or patchy spinal block in these patients mostly due to anatomical defect. There are many case reports giving blind spinal anaesthesia with modified paramedian approach & few other cases under C-arm guidance with variable success rate. [2] Here, we have successfully done SAB under USG guidance with

minimal intra-operative & post operative complications.

It has been used for real time needle guidance for providing central neuraxial blockade.

Preprocedural scan can be done to locate the midline and identify the required lumbar space and predict the depth of the space from the skin. [3]

This reduces multiple lumbar punctures, improves first attempt success rates and patient comfort.

Thus by gaining sufficient expertise for using ultrasound to guide spinal needles, the difficulty in administering central neuraxial blockade can be reduced compared to landmark technique. [3]

References

1. Castellano JM, Silvay G, Castillo JG. Marfan Syndrome: Clinical, Surgical, and Anesthetic Considerations. *Semin Cardiothoracic Vasc Anesth.* 2014 Sep;18(3):260-71.
2. Kaur M, Aujla KS, Gosal JS. Anesthetic Challenges in a Patient with Severe Thoracolumbar Kyphoscoliosis. *Anesth Essays Res.* 2020 Jan-Mar;14(1):170-172
3. Chin KJ, Karmakar MK, Peng P. Ultrasonography of the adult thoracic and lumbar spine for central neuraxialblockade. *Anesthesiology.* 2011 Jun;114(6):1459-85.