

# Thoracic segmental spinal anaesthesia for laparoscopic cholecystectomy in a case of birt-hogg-dube syndrome



Tushar Kanti Ghosh<sup>1</sup>, Purbasha Roy<sup>2</sup>

<sup>1</sup>Senior Consultant, Department of Anaesthesiology, Manipal Hospital, Kolkata, West Bengal, India, <sup>2</sup>Senior Registrar, Department of Anaesthesiology, Manipal Hospital, Kolkata, West Bengal, India

Submission: 17-07-2022

Revision: 04-02-2023

Publication: 01-03-2023

## ABSTRACT

Birt-Hogg-Dube syndrome is an extremely rare genetic disorder featuring multiple lung cyst or bulla which lead to recurrent spontaneous pneumothorax. Laparoscopic cholecystectomy poses a significant challenge in such cases from anesthetic point of view. Thoracic segmental spinal anesthesia (TSS) is a relatively new and beneficial alternative to general anesthesia to avoid post-operative complications. In this report, we used thoracic segmental spinal successfully in a known case of Birt-Hogg-Dube syndrome to avoid post-operative pneumothorax and discussed the technique and anatomical and physiological consequences. We can conclude that thoracic segmental spinal anesthesia can be used safely and successfully in laparoscopic cholecystectomy specially in cases where general anesthesia is better avoided as in Birt-Hogg-Dube syndrome.

**Key words:** Birt-hogg-dube Syndrome; Laparoscopy; Pneumothorax; Thoracic segmental spinal

### Access this article online

**Website:**

<http://nepjol.info/index.php/AJMS>

**DOI:** 10.3126/ajms.v14i3.46763

**E-ISSN:** 2091-0576

**P-ISSN:** 2467-9100

Copyright (c) 2023 Asian Journal of Medical Sciences



This work is licensed under a Creative Commons Attribution-NonCommercial 4.0 International License.

## INTRODUCTION

Laparoscopic cholecystectomy is usually performed under general anesthesia all over the world. Regional techniques such as thoracic segmental spinal and epidural anesthesia are recently emerging as a safer alternative specially in patients with chronic lung conditions to avoid post-operative pulmonary complications and prolonged hospital stay. Birt-Hogg-Dube syndrome is an extremely rare autosomal dominant disorder with a prevalence of one in 2,00,000. It is associated with three clinical manifestations including skin fibrofolliculoma, multiple lung cysts (Figure 1) and kidney tumors. These symptoms generally do not appear till adult. It is definitively confirmed by molecular genetic testing to detect frame shift mutation in folliculin (FLCN) gene on chromosome 17.<sup>1</sup> Most serious complications are

spontaneous and recurrent pneumothorax and renal cell carcinoma. Undiagnosed lung cysts and bullae may cause life threatening consequences under general anesthesia.

## CASE REPORT

The patient was a 50 years old, hypertensive, male (weight 100 kg, height 182 cm) having Birt-Hogg-Dube syndrome posted for laparoscopic cholecystectomy. Patient had history of ICU admission twice due to spontaneous pneumothorax while traveling to high altitude and had undergone video-assisted thoracoscopy for pleurodesis in the right lung few years back. Patients mother and sister also have similar condition and mother also developed pneumothorax in the immediate post-operative period after laparoscopic cholecystectomy under general anesthesia. Patients HRCT

### Address for Correspondence:

Dr. Purbasha Roy, Senior Registrar, Department of Anaesthesiology, Manipal Hospital, Kolkata, Kolkata, West Bengal, India.

**Mobile:** +91-8902445581. **E-mail:** roypurbasha1@gmail.com

thorax showed multiple moderate to large lung cyst/bulla in both lungs predominantly at bases (Figure 2). Patient was clinically stable with a METs score >6 and other pre-operative investigations were within normal limits.

After discussing with patient and surgeon, we decided to proceed with combined thoracic spinal and epidural technique as our mode of anesthesia. Inj. Midazolam 1mg and Inj. Ondansetron 4 mg were given as premedication. Normal saline 0.9% was started at 10 mL/kg body weight. Procedure was performed in sitting position. Epidural needle (18G Tuohy needle) was inserted at T9-T10 intervertebral space by paramedian approach. Catheter introduced and fixed at 9 cm. Spinal needle (26G Quincke needle) was inserted at T10-T11 intervertebral space by paramedian technique without any discomfort or paresthesia. After confirming free flow of CSF, 10 mg of Inj. Levobupivacaine 0.5% plus Inj. Fentanyl 25 mcg was injected and the patient was made supine. Sensory block was achieved from T4 to L2 dermatomes within 5 min with little weakness in the lower limbs. Oxygen was given with facemask at 4L/m and the surgeon proceeded with surgery. Inj. Midazolam 1mg, Inj. Fentanyl 50 mcg, and Inj. Ketamine 25 mg was given intravenously before CO<sub>2</sub> insufflation. Patient did not require any further sedation throughout the surgery. Intra-abdominal pressure was kept between 10 and 12 mm Hg. Surgery continued uneventfully for 60 min. Drop in blood pressure from 152/90 mm Hg to 92/56 mm Hg was managed with Inj. Mephentermin 6 mg iv. Heart rate was maintained between 52–60/min throughout the surgery. Total 1500 mL of crystalloid was given in perioperative period. Activation of epidural route was not required during surgery. Inj. Levobupivacaine 0.125% 8 mL plus Inj. Fentanyl 50 mcg was given epidurally 10 min before ending of surgery for postoperative analgesia. Recovery was uneventful without any neurological sequelae or pneumothorax or PONV or urinary retention. Analgesic requirement was minimum for 1<sup>st</sup> 24 h and the patient was discharged on day 3 without any complications.

## DISCUSSION

Administration of anesthesia in a case of Birt-Hogg-Dube syndrome is extremely challenging. Positive pressure ventilation and PEEP during general anesthesia itself may lead to rupture of bulla causing pneumothorax. In addition to that raised intra-abdominal pressure due to pneumoperitoneum and trendelenburg position during laparoscopic cholecystectomy may increase risk further more. Kumasaka et al.,<sup>2</sup> believed that the loss of FLCN leads to the weakening of the alveolar wall, which is easily

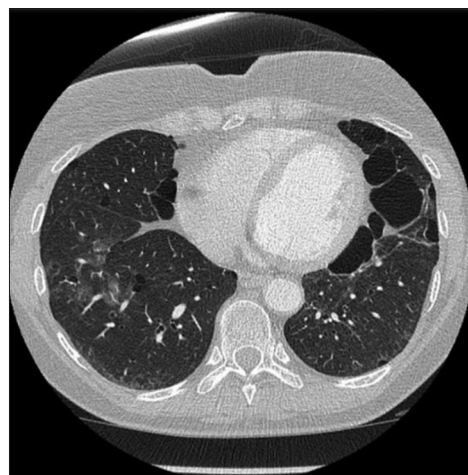


Figure 1: Axial view of CT thorax of a case of Birt-Hogg-Dube syndrome<sup>9</sup>

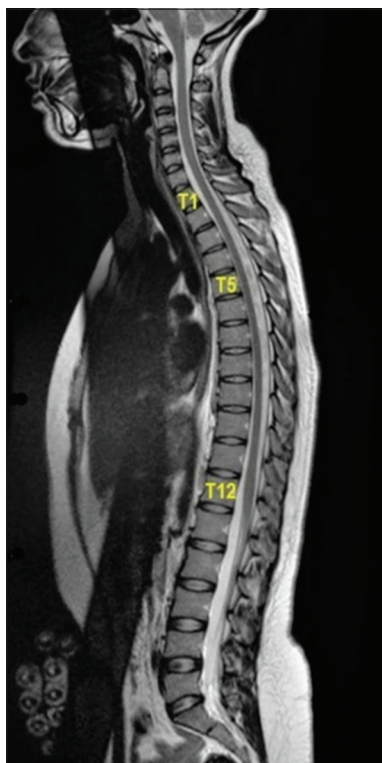


Figure 2: CXR PA view of patient

destroyed by mechanical pressure during respiration, which leads to cystic changes.

TSS anesthesia is a newly evolving topic which involves injection of small volume of preservative free isobaric local anesthetic drug like levobupivacaine in thoracic subarachnoid space. It raises three major concerns: Neural injury and its sequelae, hemodynamic changes due to high level of sympathetic block, and respiratory distress due to motor block of accessory muscles of respiration.

Imbelloni et al.,<sup>3</sup> investigated the anatomy of thoracic spine and postulated that the space between the dura matter and spinal cord in the thoracic region measured with MRI was 5.19 mm at T2, 7.75 mm at T5, and 5.88 mm at T10 (Figure 3). The lumbar spinal cord is situated more dorsally and takes up more space because of the lumbar enlargement so that it is at greater risk of needle damage as shown by Reynolds's reports of pain and paresthesia when needles were inserted at that level.<sup>4</sup>



**Figure 3:** T2-weighted sagittal MRI of a normal spine<sup>10</sup>

Bradycardia and hypotension during TSS are mainly attributed to inhibition of cardioaccelerator fibers (T1-T4) and sympathetic block due to thoracolumbar distribution of the drug. A study comparing anesthetic techniques of laparoscopic cholecystectomy showed that TSS anesthesia was better than lumbar spinal, regarding hemodynamic stability (lower dose of ephedrine).<sup>5</sup> It may be due to use of low dose isobaric local anesthetic in thoracic level and blockade of fewer dermatomes sparing lower limbs than lumbar spinal anesthesia. The thoracic nerve roots are thinner compared with lumbar nerve roots with reduced cerebrospinal fluid volume at this level, allowing for more efficient nerve blockade as there is less dilution of the LA agent; thus a lower volume of LA is needed for adequate nerve blockade.<sup>6</sup>

Respiratory distress was not there as the main respiratory muscle diaphragm is innervated from cervical level and thus spared during thoracic spinal. Expiration is a passive process (does not normally require accessory muscles of respiration), is not normally affected by the block. However, in patients with chronic lung disease, who use anterior abdominal muscles (innervated from thoracic level) for expiration, ventilatory impairment may happen.<sup>7</sup> It may be possible that use of low volume of drug reduced the degree of motor block. Intra-abdominal pressure was kept low to allow adequate diaphragmatic excursion.

We used isobaric levobupivacaine which is a S-enantiomer of bupivacaine, also caused early regression of motor

block. Early administration of sedatives such as midazolam, fentanyl, and ketamine before CO<sub>2</sub> insufflation avoided patient discomfort due to shoulder tip pain.

Zundert et al.,<sup>7</sup> also used TSS successfully for laparoscopic cholecystectomy in patients with severe lung diseases. Madishetti et al.,<sup>8</sup> even used it for mastectomy.

We kept thoracic epidural catheter in place as a back-up in case of failure of TSS as we lack skill because it is still a less practiced technique in our hospital and we certainly tried to avoid general anesthesia in this case at all cost. Later, we used epidural for post-operative analgesia which also increased patient satisfaction.

Every patient responds differently to anesthesia which makes every case unique. There is never a right way to anesthetize a patient. The idea is to provide best care according to personal choice and different expertise. Thus, we chose TSS as our technique and we were able to avoid lung complications of general anesthesia and hemodynamic instability of lumbar spinal anesthesia in this particular case (Birt-Hogg-Dube syndrome).

## CONCLUSION

In the end, we can conclude that thoracic segmental spinal anesthesia can be used safely and successfully in laparoscopic cholecystectomy specially in cases where general anesthesia is better avoided as in Birt-Hogg Dube syndrome. It can also be used routinely but will require further studies involving large sample size to enforce that.

## REFERENCES

1. Lee JH, Jeon MJ, Song JS, Chae EJ and Choi JH. Birth-Hogg Dubé syndrome in Korean: Clinicoradiologic features and long term follow-up. *Korean J Intern Med.* 2019;34(4):830-840. <https://doi.org/10.3904/kjim.2018.119>
2. Kumasaka T, Hayashi T, Mitani K, Kataoka H, Kikkawa M, Tobino K, et al. Characterization of pulmonary cysts in Birt-Hogg Dube syndrome: Histopathological and morphometric analysis of 229 pulmonary cysts from 50 unrelated patients. *Histopathology.* 2014;65(1):100-110. <https://doi.org/10.1111/his.12368>
3. Imbelloni LE, Quirici MB, Filho JR, Cordeiro JA and Ganem EM. The anatomy of the thoracic spinal canal investigated with magnetic resonance imaging. *Anesth Analg.* 2010;110(5):1494-1495. <https://doi.org/10.1213/ANE.0b013e3181d5aca6>
4. Reynolds F. Damage to the conus medullaris following spinal anaesthesia. *Anaesthesia.* 2001;56(3):238-247. <https://doi.org/10.1046/j.1365-2044.2001.01422-2.x>
5. Yousef GT and Lasheen AE. General anesthesia versus segmental thoracic or conventional lumbar spinal anesthesia for patients undergoing laparoscopic cholecystectomy. *Anesth Essays Res.* 2012;6(2):167-173.

- <https://doi.org/10.4103/0259-1162.108302>
6. Imbelloni LE and Gouveia MA. A comparison of thoracic spinal anesthesia with low-dose isobaric and low-dose hyperbaric bupivacaine for orthopedic surgery: A randomized controlled trial. *Anesth Essays Res.* 2014;8(1):26-31. <https://doi.org/10.4103/0259-1162.128900>
  7. Van Zundert AA, Stultiens G, Jakimowicz JJ, van den Borne BE, van der Ham WG and Wildsmith JA. Segmental spinal anaesthesia for cholecystectomy in a patient with severe lung disease. *Br J Anaesth.* 2006;96(4):464-466. <https://doi.org/10.1093/bja/ael036>
  8. Madishetti ER, Aasim SA, Bethi R. Segmental thoracic spinal anaesthesia for modified radical mastectomy in a bronchiectasis patient. *Indian J Anaesth Anal.* 2017;4(2):546-550. [https://doi.org/10.21088/ijaa.2349.8471.42\(pt-II\)17.31](https://doi.org/10.21088/ijaa.2349.8471.42(pt-II)17.31)
  9. Glick Y. Birt-Hogg-Dube Syndrome. Case Study. *Radiopaedia. Org.* Available from: <https://radiopaedia.org/cases/91272/studies/108876?lang=us> [Last accessed on 2023 Jan 09].
  10. Ibrahim D. Normal MRI Spine. Case Study. *Radiopaedia. org.* Available from: <https://radiopaedia.org/cases/92846> [Last accessed on 2023 Jan 09].

**Authors' Contributions:**

**TKG**- Review of manuscript and treating consultant anesthesiologist and **PR**- Concept and design, manuscript preparation, revision of manuscript, and treating anesthesiology registrar.

**Work attributed to:**

Manipal Hospital, Kolkata-700106, West Bengal, India.

**Orcid ID:**

Dr. Tushar Kanti Ghosh - <https://orcid.org/0000-0002-0256-9239>

Dr. Purbasha Roy - <https://orcid.org/0000-0003-3851-0674>

**Source of Support:** Nil, **Conflicts of Interest:** None declared.