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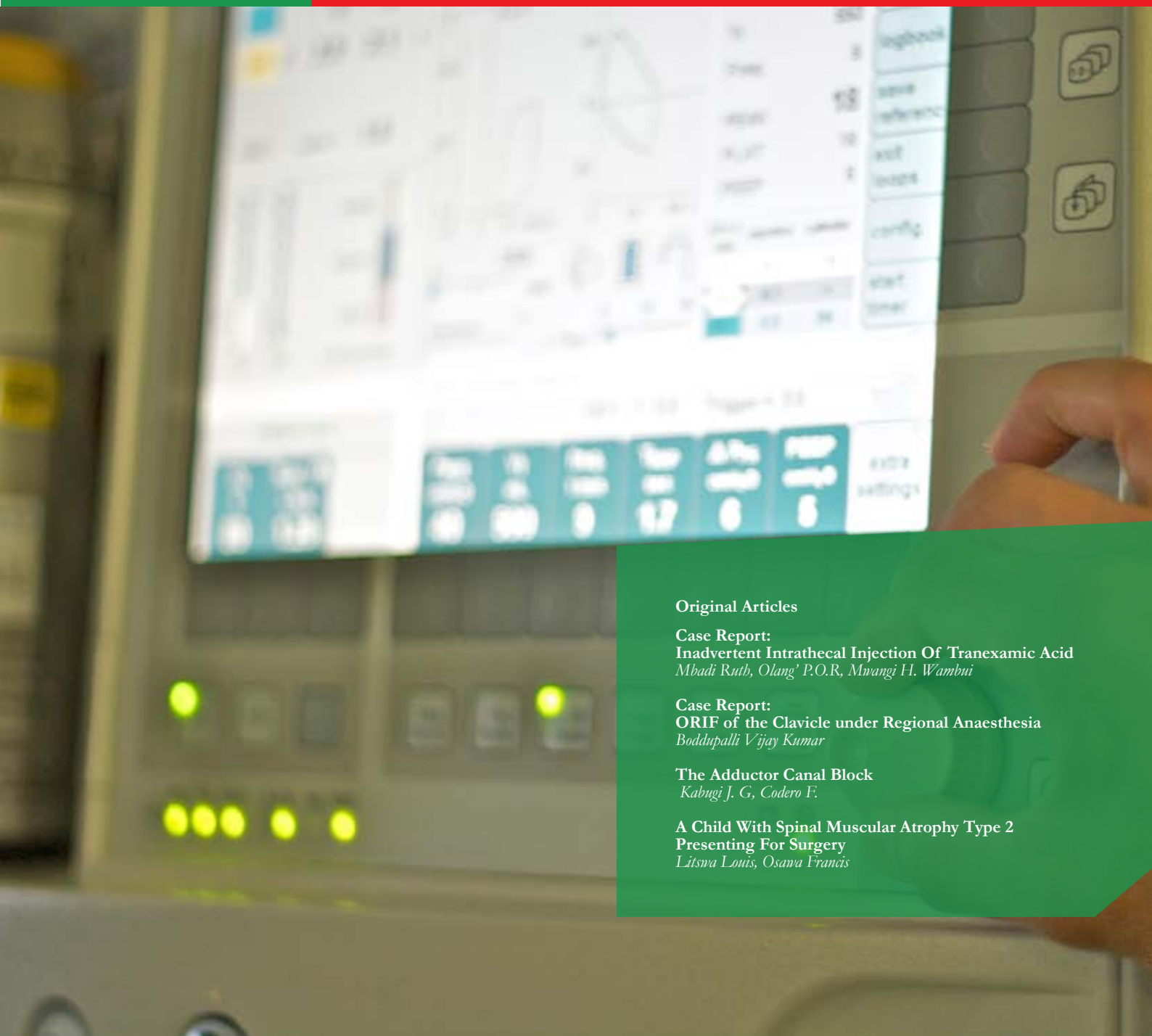

Original Articles

Case Report:
Inadvertent Intrathecal Injection Of Tranexamic Acid
Mbadi Ruth, Olang' P.O.R, Mwangi H. Wambui

Case Report:
ORIF of the Clavicle under Regional Anaesthesia
Boddupalli Vijay Kumar

The Adductor Canal Block
Kabugi J. G, Codero F.

A Child With Spinal Muscular Atrophy Type 2 Presenting For Surgery
Litswa Louis, Osawa Francis

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- Title
- Abstract
- Introduction
- Methods
- Results
- Discussions
- Conclusion
- References

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Editorial

The practice of Anaesthesiology is unique in demanding almost consistently the immediacy of apt, demonstrable and appropriately reversible pharmacologic activity of clinically applicable reactants. The review, prescription, administration and management of regular anaesthetic agents is routinely a primary undertaking and responsibility of the attending anaesthesiologists in clinical circumstances where anaesthetic services are provided. The need for administration of multiple agents concurrently and in quick succession requires strict vigilance of these highly potent agents in use as well as the patient's response not only to the surgical and disease insult but the potential drug effects and possible interactions.

Research and development has deployed to the anaesthesiologist an ever-increasing armamentarium of pharmacological agents in the perioperative space. It is also not lost to the many suppliers and manufacturers what this demand for drugs portend in economics of both health care and financial returns. Increasingly therefore with limited universal guidelines and probity of enforcement, the manufacture and sale of drugs that bear similar packaging and names is becoming a major concern in clinical practice.

There is an inherent danger for these 'look alike and sound alike' (LASA) drugs to be mistakenly administered with potentially catastrophic results. Very few agents bear unique physical characteristics to differentiate them and with the routine basically poly pharmaceutics approach to balanced anaesthesia; the practice of Anaesthesiology thus lends itself easily to potential and actual errors in terms of drug dosages as well as administration. This journal presents a case report on the inadvertent intrathecal administration of tranexamic acid for a caesarean section and outlines the events that ensued and subsequent management.

In this issue, we highlight the ever increasing use of regional anesthetic techniques in the face of trauma and limb surgery that improve perioperative outcomes: the use of cervical and interscalene blocks to enable surgical management for clavicular fractures in the face of lung contusion and difficult airway access is certainly a useful technique to be learnt as trauma remains a leading cause of morbidity and mortality in our setting.

Attention to the principles and conduct of the adductor canal block for perioperative analgesia following knee arthroplasty is equally addressed: the advantage of retained motor function with this technique results in earlier ambulation and reduced chances of the risk for a fall in this patient population.

Neuromuscular disorders present a constant concern in anaesthetic practice as they may lead to respiratory and postural challenges: respiratory challenges may result in prolonged hospital stay, dependency on mechanical ventilation with scenarios predisposing to pneumonic and atelectatic processes. Postural challenges on the other hand may lead to difficulties in conducting regional anaesthetic techniques, hinder surgical access and a predilection to decubitus ulcers.

In the last article of this edition, we present a case of spinal muscular atrophy type 2 and its successful perioperative management while taking cognizance of potential complications that may arise.

Case Report

Inadvertent Intrathecal Injection Of Tranexamic Acid

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Abstract

Medication errors are a common cause of morbidity and mortality in medicine. Unlike most specialities, drugs used in anaesthesia have quick onset of action and the consequence of medication errors can be more serious than in most other specialities. We report an accidental intrathecal injection of tranexamic acid instead of 0.5% hyperbaric bupivacaine during spinal anaesthesia for an emergency caesarean section in a 25 year old primigravida. Thirty minutes after anaesthesia administration, she developed progressive neurological symptomatology with resultant post-operative critical care admission. The patient made full recovery through the multi-organ support after 13 days. There was no report of lack of anaesthetic effect during surgery.

Introduction

The United States National Coordinating Council for Medication Error Reporting and Prevention defines a medication error as "any preventable event that may cause or lead to inappropriate medication use or patient harm while the medication is in the control of the health care professional, patient, or consumer. Such events may be related to professional practice, health care products, procedures, and systems, including prescribing, order communication, product labelling, packaging, and nomenclature, compounding, dispensing, distribution, administration, education, monitoring, and use" (1)

The factors associated with medical errors include but are not limited to healthcare staff, patients, work environment and the medicine itself (2).

Tranexamic acid is an anti-fibrinolytic agent whose use is popular in obstetric, cardiac and gynaecological procedures. Cases of inadvertent intrathecal injection of the same have been reported to have resulted in neurotoxicity manifesting in seizures and cardiotoxicity manifesting in arrhythmias. In this case, the patient developed seizures and was taken to ICU where she recovered fully and walked home with no residual neurological sequelae.

Case Description

We report a case of a 25 year old primigravida who was brought for emergency caesarean delivery due to prolonged labour. Anaesthetic evaluation was unremarkable.

2 mls of solution was injected intrathecally and the surgery started. There was no report of inadequate anaesthesia. During extraction of the baby, the mother reported that she; "felt funny in the back and legs". Shortly after, she developed "un-intentional, intermittent jerking of the legs". These then spread to the upper limbs at which point she became unresponsive and developed generalised convulsions.

Diazepam 10mg was given intravenously, and additional 50mg intramuscular phenobarbitone (normal seizure control dose is 20mg/kg slow i.v. push). With little control of the the jerky movements to the fetal extraction; a live female infant with an Apgar score reported as 8/1 and 9/5.

Ventimak oxygen supplementation was substituted fir endotracheal intubation and mechanical ventilation with significant intraoperative tachycardia at 130-150 bmp from pulse oxymetric tracing. ECG rhythm was not available since the pulse readings were from a pulse oximeter. Only one reading of non invasive blood pressure of 68/42mmHg was recordable during the convulsive and resuscitative phase.

Critical evaluation on medication administration on the anaesthetic revealed an unusual ampoule had been opened for the spinal anaesthetic, which was actually 500mg tranexamic acid and not the intended 10mg bupivacaine.

Post operatively, the patient was transferred to the critical care unit and induced into a barbiturate coma for convulsion control for 24 hours. She required up to 300mg per hour for optimal convulsive management. There was a prolonged comatose phase on termination of the sodium thiopentone infusion but no recurrence of the convulsions and regularisation of sinus cardiac rhythm.

Her renal function however deteriorated with initial fluid retention and significant derangement of renal biochemical markers necessitating institution of support. Haemodialysis was commenced on the fifth day post operatively with significant improvement. She regained consciousness gradually after three sessions on dialysis.

On day 13, she was fit for transfer to regular ward care and finally was discharged home with no renal or neurological sequelae.

Discussion and Conclusion

Tranexamic acid is an anti-fibrinolytic medication which has been shown in the trauma literature to be highly effective in reducing deaths due to bleeding. In the WOMAN (World Maternal Antifibrinolytic) Trial Collaborators (Lancet, 2017) 6 it was observed that death due to post-partum bleeding was reduced by approximately 30% if the drug was given within 3 hours of childbirth. The prevalence of laparotomy to control bleeding was similarly reduced significantly. It is for these, and other reasons that tranexamic acid is readily available in most labour and delivery suites as well as maternity operation rooms.

Patients who had received accidental intrathecal injection of tranexamic acid (TXA) reported severe back pain that radiated below the waist, with burning pain in the lower limbs and gluteal region, Involuntary motor activity, such as a "jerking" of the lower extremities (referred to as myoclonic movements) and twitching of facial muscles, was also observed. These abnormal movements rapidly progressed to generalized tonic–clonic seizures. Studies show that TXA directly increases the excitability of neuronal networks. Increasing evidence suggests that this hyperexcitability produced by TXA results from reduced inhibitory neurotransmission or "disinhibition." Aminobutyric acid type A (GABAA) receptors and glycine receptors are major mediators of inhibition in the CNS(3). Anaesthetic agents such as propofol and sodium thiopentone (STP) have been used in some cases to control the seizures. The pathophysiology of the cardiac manifestations is not well understood.

A summary of the reported cases of accidental intrathecal injection of tranexamic acid that have been recorded is attached below.

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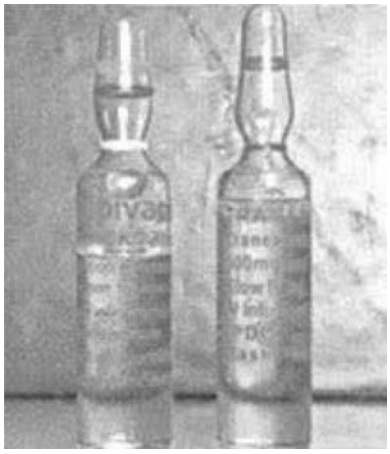

The drug error reported above occurred largely due to labelling and packaging of similar ampoules among other reasons like workload and work pressure due to the nature of the case being an emergency and probably poor communication among the team members.

The delay in commencement of the dialysis for this patient was occasioned by a delay in obtaining necessary investigations requested for by the renal team further outlining the challenges faced in areas where resources maybe limited and not easily obtainable in the critical care set up. Her delayed emergence could be a combined factor of both the barbiturate load as well as the incipient renal dysfunction.

With regards to the anaesthesia, the patient apparently was well anesthetized for the surgical incision and manipulations. In the first reported case of intrathecal tranexamic acid in the 1970s, the surgery was done successfully without the patient feeling any pain.

Jensen et al developed strategies for preventing drug administration errors during anaesthesia from a systematic review of literature (4). One general and five specific strong recommendations were generated: systematic countermeasures should be used to decrease the number of drug administration errors in anaesthesia; the label on any drug ampoule or syringe should be read carefully before a drug is drawn up or injected; the legibility and contents of labels on ampoules and syringes should be optimised according to agreed standards; syringes should (almost) always be labelled; formal organisation of drug drawers and workspaces should be used; labels should be checked with a second person or a device before a drug is drawn up or administer.

Since 2010, each hospital in the US is required to comply with Medication Management Standard by developing a list of look-alike/sound-alike medications. The hospital develops a list of look-alike/sound-alike medications it stores, dispenses, or administers. One source of look-alike/sound-alike medications is The Institute for Safe Medication Practice's.

(ISMP's) List of Confused Drug Names. It contains look-alike and sound-alike (LASA) name pairs, of medications that have been published in the ISMP Medication Safety Alert!® and the ISMP Medication Safety Alert!® Community/Ambulatory Care Edition.

This list is used to determine which medications require special safeguards to reduce the risk of errors and minimize harm. It may include strategies such as:

i. Using both the brand and generic names on prescriptions and labels.

ii. Including the purpose of the medication on prescriptions.

iii. Configuring computer selection screens to prevent look-alike names from appearing consecutively.

iv. Changing the appearance of look-alike product names to draw attention to their dissimilarities.

v. Both the FDA-approved and the ISMP-recommended tall man (mixed case) letters have been included in this list.

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Case Report

ORIF of the Clavicle under Regional Anaesthesia

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Abstract

Blunt chest trauma, depending on the impact, may result into fracture of Clavicle, fracture of ribs and associated lung contusion. The extent and severity of lung contusion may result in a ventilatory perfusion mismatch with subsequent hypoxemia manifesting a lower than optimum oxygen saturation (Spo2). In some of these cases, conservative medical management may be adopted with analgesics and supplementary oxygen administered by mask provided the basic respiratory function is maintained. A proportion of patients with clavicle fractures may, however, need open reduction and internal fixation (ORIF) of the clavicle with Regional Anaesthesia i.e. Interscalene Brachial Plexus Block + Superficial Cervical Plexus Block offering a probable better outcome than general anaesthesia in view of the postoperative morbidity - respiratory dysfunction (hypoxia and hypercarbia) and a need for postoperative mechanical ventilation.

Introduction

Blunt chest trauma is common in motor vehicle accidents, falls and sports. The incident rate of blunt chest trauma is high in productive age group.

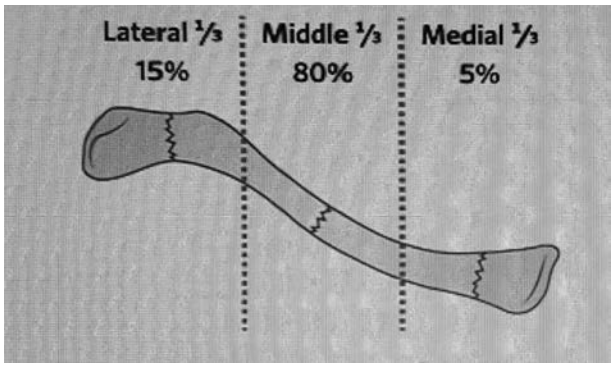


Fig: 1. Classification of Clavicle Fractures by Location:
1. Lateral--15%
2. Middle--80%
3. Medial--5%

	No. of pts.	2 or >2 Ribs#	Multiple Ribs 3 or>3#
Pulmonary Contusion	139	15	12
Pulmonary Laceration	3	0	3
Pneumothorax	40	10	30
Haemothorax	45	5	40
Haemo pneumothorax	25	3	22
*Fracture Clavicle	11	1	10
Sternal Fracture	6	2	4
Scapula Fracture	5	0	5

Courtesy: Trakia University; Bulgaria 2017.

Fig; 2. In Blunt Chest Trauma: Associated Ribs Fracture and Chest injuries according to the type of Ribs Fractures.

Case Report

An 85kg 63 years old male presented to hospital with a fracture of the right side middle clavicle. The patient sustained blunt chest trauma due to head on collision of his moving motor vehicle with another motor car on the 12th September 2018.

Chest X-ray: Right clavicle middle comminuted fracture.
Right lower lobe Contusion/consolidation.
Right side fractures of the first to sixth ribs.

Patient was admitted into ICU. He was dyspnoeic, pulse rate 100/min, spo2 on room air hovering around 84%, BP 140/80 mmHg, Resp. rate 20 to 22/min. Patient was conservatively managed with O2 mask, analgesics and the beach chair position for 2 days. On the 14th of September, the vital signs were: Pulse rate 80/min, BP 137/77 mmHg, Resp. Rate around 22/min., Spo2 on room air ranging 84% to 86%, and rising to 95% on 5 liters of oxygen by mask. Examination of the cardiovascular and central nervous system were unremarkable.

Airway examination revealed a receding mandible with an anterior placed larynx (difficult intubation). Air entry on the right side of the chest was reduced on auscultation.

Problems

Difficult Airway-difficult Intubation, Spo2-on room air-84% to 86%.

A decision to perform ORIF on the clavicle under regional anaesthesia was made: Ultrasound guided Interscalene Brachial Plexus Block + Superficial Cervical Plexus Block. The patient lay on the table in Semi-Fowler's position at 30 degree with his face to the left side. O2 mask with 5 lit/min was administered, Inj. midazolam 1mg. & inj. Fentanyl 50 +25 mcgs given intravenously in titration with spo2. Regional anaesthetic solution was prepared: 12.5ml of 0.5% Bupivacaine + 12.5ml of 2% xylocaine with adrenaline.

Interscalene Block

Under ultrasound guidance , a Linear Probe(high frequency) was used to locate the plexus, a 22G spinal needle was inserted in posterior approach and 15ml of the prepared local anaesthetic solution was injected around the plexus(signal light area) aseptically.

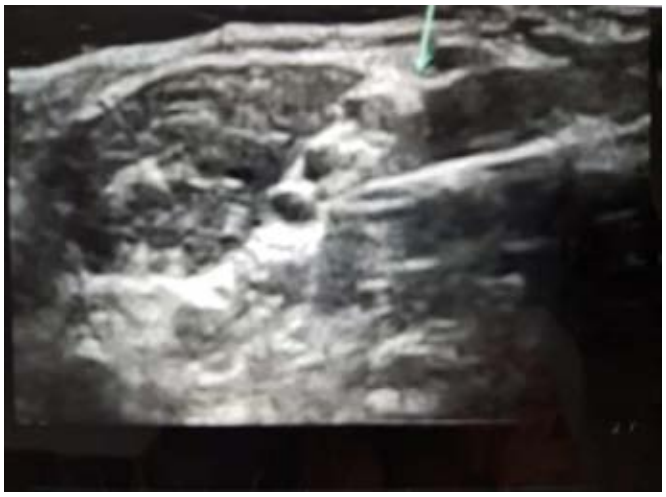
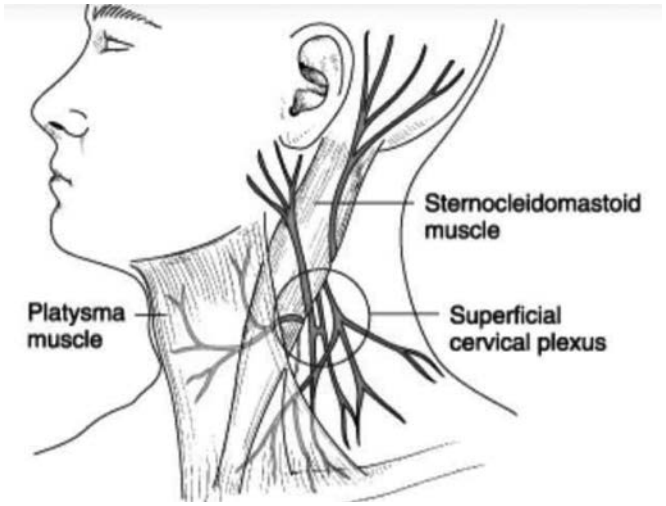
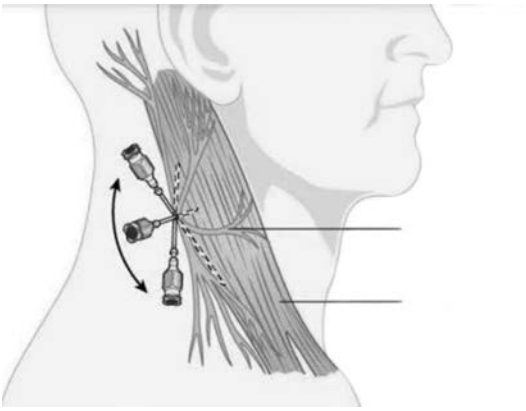


Fig: 3. Interscalene block-Needle pointing to the Traffic signal sign.

Superficial Cervical Plexus Block: It was performed as plane block with 10ml of the prepared regional anaesthetic solution injected through a marked point in fan shape. The marked point was a line was drawn from the tip of the mastoid process along the posterior border of the clavicular head of the SCM with the midpoint of the line i.e. where the external jugular vein crosses was the point of needle insertion. 25G, 1 " needle was inserted 1 to 1.5cm depth behind the SCM to avoid the deeper block.





Superficial Cervical Plexus Block: It was performed as plane block with 10ml of the prepared regional anaesthetic solution injected through a marked point in fan shape. The marked point was a line was drawn from the tip of the mastoid process along the posterior border of the clavicular head of the SCM with the midpoint of the line i.e. where the external jugular vein crosses was the point of needle insertion. 25G, 1 " needle was inserted 1 to 1.5cm depth behind the SCM to avoid the deeper block.

Onset of action of the block was within 10minutes and clavicle surgery commenced (plate and screws fixation) and completed successfully in an hour without any complaint of pain or discomfort from the patient. Vital sings remained stable through the procedure with little variation: pulse rate 80+or- 10/min, BP 130/77 + or - 7 mmHg, SpO2 on O2 mask 5lit/min 93 +or- 3%. There was no necessity of any additional analgesics intraoperatively.

Effective analgesia as reported by the patient lasted approximately 150 minutes and tramadol and paracetamol were subsequently utilized for postoperative analgesia.

15.09.2018: 1st postoperative day or 72 hours after accident SpO2...77% on room air. CT chest confirmed the Right lung lower lobe contusion.

16.09.2018: 2nd postoperative day or 4 days after accident SpO2...88% on room air & 94% with O2 mask.

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The Adductor Canal Block

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2. Fellow Neuroanaesthesia

Abstract

Saphenous nerve blockade provides reliable and profound analgesia devoid of lower limb muscle weakness especially for knee surgery. The Adductor Canal approach is one approach that selectively provides this advantage in a simplified and safe access. This is remarkable especially for anterior knee skin surgical approach. Regular medial aspect surgical procedures such as Saphenous Vein harvesting for CABG or wound suturing are adequately covered by this peripheral nerve block approach. Quadriceps weakness seen with the Femoral Nerve Block doesn't occur with this profile.

Introduction

Enteral and parenteral multimodal analgesic management has been the clinical approach to lower limb surgery. Total Knee Arthroplasty pain can additionally be managed with augmentation of peripheral nerve blockade. The femoral nerve is the main motor and sensory innervation of the affected surgical field. The nerve block technique was initially a Femoral Nerve Block (FNB) or

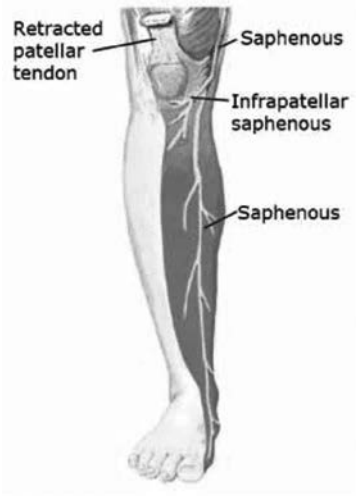
Proximal femoral nerve block (FNB) is simple to perform. It has easily outlined landmarks (Femoral Artery and Vein) but has the drawback of quadriceps weakness. This is significant in that motor weakness has resulted in falls that put hips at risk of fracture with delayed mobilization and healing. The Adductor Canal Block (ACB) is essentially a block targeting the saphenous branch of the femoral complex. The ACB is a deeper block but preserves quadriceps strength since only the Nerve to Vastus Medialis is blocked. Being technically easier therefore makes the ACB a better option for knee surgery.

Indications for the adductor canal block

- Knee surgery including arthroplasty, Patella and Patella tendon surgery
- Saphenous vein harvest for CABG (Coronary angio bypass graft surgery)
- Together with Popliteal Sciatic Nerve Block, it provides for anesthesia/analgesia at and below the knee

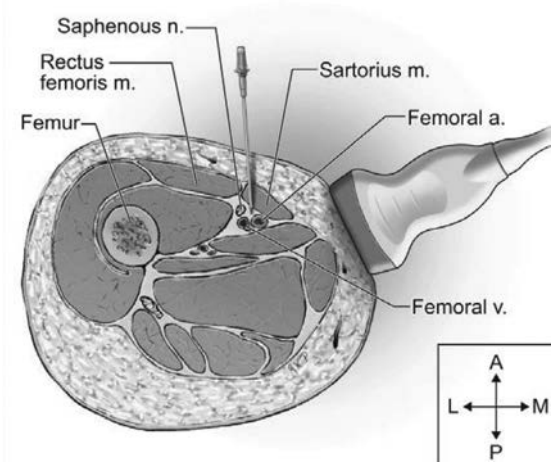
Anatomy

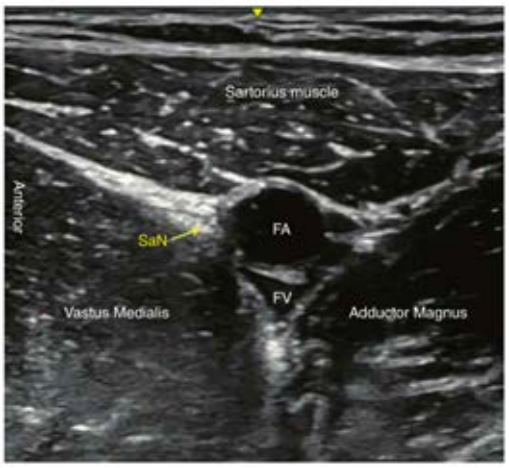
The Saphenous Nerve is the terminal sensory branch of the Femoral Nerve. It is contained in the subsatorial or Hunters canal in the anterior aspect of the thigh. It gives off infrapatellar branches that supply the knee joint and its anterior surface which is the regular skin incision and surgical access for Knee arthroplasty. It then goes on to provide sensory supply to the medial side of the leg, ankle and hindfoot.



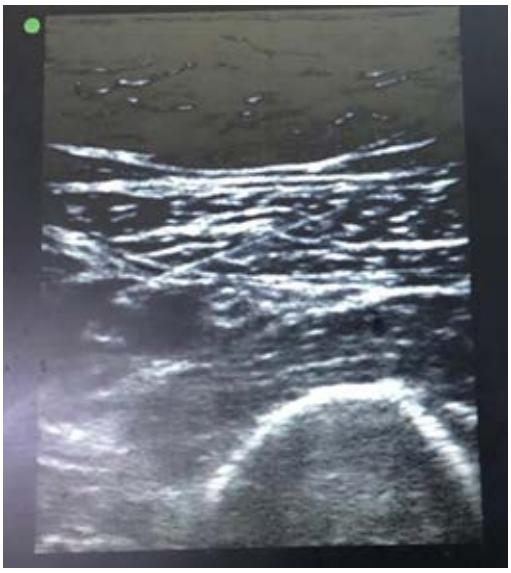
Technique and Sonographic Anatomy

The aim is to inject the local anesthetic mixture in Hunter's Canal which is bounded by the Sartorius anteriorly, Vastus Medialis (laterally) and Adductors Magnus and Longus (medially). It contains The Femoral vessels, The Saphenous Nerve and The Nerve to Vastus Medialis. The nerve to the Vastus Medialis is motor in nature and the only quadriceps muscle affected when the ADC block is applied preserving the muscle power of the other knee extensors.





The sonographic anatomy of the adductor canal.



The needle is approached the Femoral Artery in an anteroposterior orientation and the drug is injected to surround the artery while avoiding intravascular injections.

Block Performance

In an aseptic approach the block is performed using A 100 mm Stimulating nerve block needle, with application of 25-30 mL of Local Anesthetic mixture: Bupivacaine 0.2% with or without adrenaline 1:250000 or Bupivacaine with or without dexmedetomidine 0.025-0.1 mcg/kg. The high frequency linear Ultra Sound probe aids in localization of injection area. It is placed on the medial aspect of the mid-thigh to visualize the femoral artery under the Sartorius muscle. This has been likened to a boat on a log. The needle is then advanced through the Sartorius muscle to lie near the femoral artery and the injection made ensuring that the drug surrounds the artery. Several needle passes may be required to ensure this. A negative for blood aspiration is paramount before drug administration since two large vessels, the femoral artery and vein, lie in this canal.

Conclusion

The ACB is favored over the FNB since it blocks only the Saphenous Nerve and the Nerve to Vastus Medialis leaving the rest of the quadriceps intact. The clinical implication of this is that there is very little to no muscle weakness with the ACB as opposed to the FNB which results in quadriceps paralysis predisposing the patient to dangerous falls. It is also very easy to learn and perform since the femoral artery is an easy landmark. The infrapatellar branches may be theoretically used to only provide analgesia for the knee without the medial leg numbness that occurs with the ACB.

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A Child With Spinal Muscular Atrophy Type 2 Presenting For Surgery

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Introduction

Spinal muscular atrophy type 2 is a rare autosomal recessive disease characterized by muscle wasting and may often result in death due to complications arising from respiratory muscle compromise. The muscle wasting leads to increased sensitivity to neuromuscular junction blockers as well as postural anomalies that may resulting in various anaesthetic challenges affecting securing of the airway, ventilation and patient positioning.

Patients with muscular spinal atrophy type 2 may present for corrective spine surgery or other surgical interventions aimed at eventual spine surgery: in this instance, the patient was scheduled to undergo a feeding gastrostomy to optimize her nutritional status prior to spine surgery for scoliosis.

Case Presentation

BWK, a thirteen year old girl, presented for insertion of a feeding gastrostomy under general anaesthesia intended to improve her nutritional status to optimize her for subsequent vertebral corrective surgery for scoliosis. This was her index anaesthetic exposure.

The diagnosis of spinal muscular atrophy had been made at the age of two by genetic and muscular testing following delay in onset of the usual motor milestones and recurrent respiratory tract infections. Management for her condition was supportive and she was wheelchair bound. There was, however, no familial history of similar disease.

History taking elicited a complaint of constipation managed on laxatives, whilst physical examination revealed an alert intelligent girl severely wasted at 14 kilograms with severe cervicothoracic scoliosis and tremors of the limbs at rest. No pressure sores were noted. There was a 'bell' like appearance of the thorax with reduced air entry over the left side though the oxygen saturation on room air was 92% suggesting probable long-term compensation as the breathing rate was 16 breaths per minute without signs of distress. The abdomen was centrally scaphoid, had a firm nonballotable right hypochondrial mass but no tenderness. The lower limbs were flexed at the hip and knees due to contractures. The pulse and blood pressure indices were normal for age.

Assessment of the airway revealed a Mallampati class 4 with an interincisor gap of 3 inches and a sternomental distance of 6 centimeters with a head rotated and fixed at about 20 degrees to the right.

A chest x-ray ordered revealed a large gas shadow beneath the diaphragm with reduced left lung markings and severe kyphoscoliosis (fig 1).

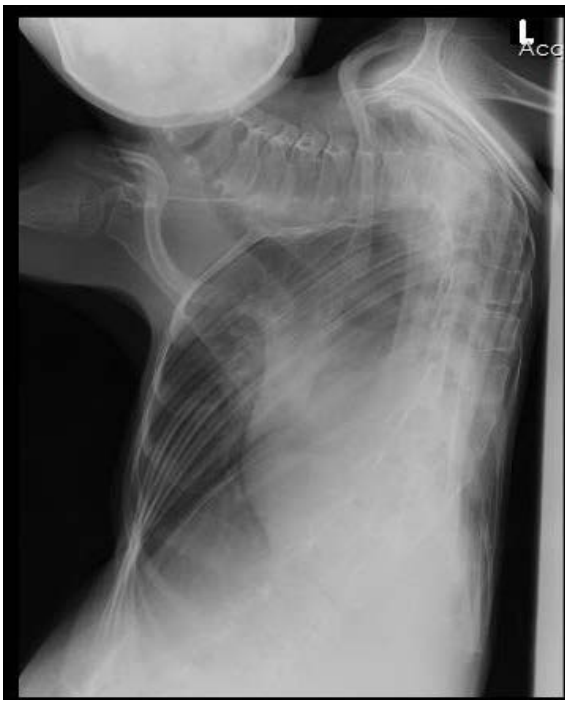

Fig1: Note the severe scoliosis, the bell shaped chest and large gastric air bubble.

A decision to proceed with surgery was made and induction was by 1.5mg of midazolam, 50mg of propofol and an infusion of remifentanil starting at 0.2 micrograms per kilogram with 100% oxygen. An assessment of the airway revealed an anterior laryngeal inlet precede by grade 4 tonsillar hypertrophy necessitating use of atracurium at 0.3mg/kg to enable intubation and ideal surgical access. Intubation was successful on the third attempt using a size 5.5 uncuffed oral endotracheal tube and a size 3 flexible tip McCoy laryngoscope obviating a need for videolaryngoscopy.

Intraoperative analgesia was provided by 375mg of paracetamol and 12.5mg of diclofenac rectally as well as incisional 2mg/kg bupivacaine with maintenance of anaesthesia on propofol at 60 micrograms /kg/minute, remifentanil at 0.1 micrograms/kg/minute and a mixture of 50% oxygen in air. Surgical access revealed gaseous gastric distension probably explaining the x-ray findings. Vital signs and oxygenation remained normal with an uneventful procedure lasting for 45 minutes and no further neuromuscular blockade was required. Reversal of neuromuscular blockade using 0.05mg/kg neostigmine and

0.02 mg/kg atropine as well as discontinuing the propofol and remifentanil infusions resulted in wakefulness in a painless child with return of adequate spontaneous respiration.

Postoperative analgesia was paracetamol and ibuprofen with a visual analogue score of 1-2 thus no opiate was required. The oxygen saturation remained above 93% postoperatively with a good cough reflex obviating a need for chest physiotherapy.

BWK was subsequently discharged home after 4 days.

Case Discussion

Spinal muscular atrophy (SMA) is a rare autosomal recessive hereditary disorder characterized by loss of lower motor neurons and progressive muscular wasting often leading to early death. It is all known as Autosomal Recessive proximal spinal muscular atrophy or 5q spinal muscular atrophy. It has a frequency of 1:5000-1:12000 people globally.

SMA results in motor neuronal death in both the brainstem and the spinal cord and will often result in death due to difficulties in breathing or complications of inability of motor neuronal activity such as inability to eat, postural infections, etc. Motor neurons are not only responsible for muscle activity, they are also trophic for muscle growth and maintenance hence their 'absence' leads to muscle atrophy (fig 2).

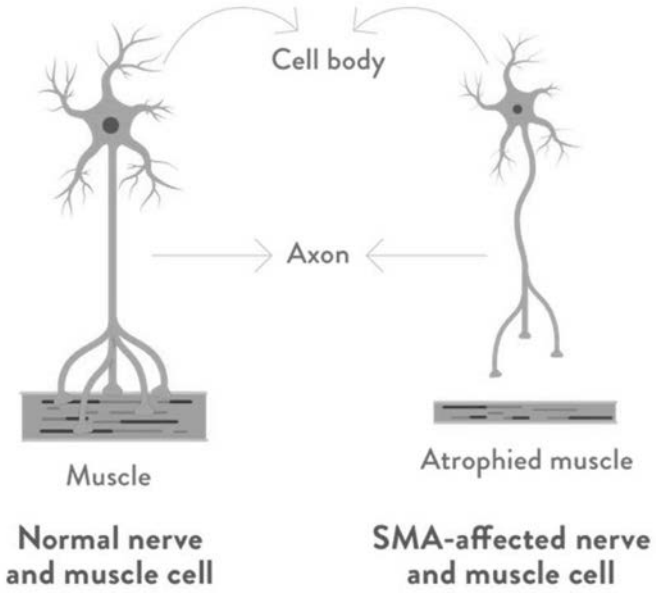

Fig 2.

SMA occurs as a result of a genetic mutation in the formation and subsequent activity of survival motor neuron proteins (SMN) coded for by the gene SMN1: failure to express these proteins may occur early in life hence SMA types 1 and 2, with the former manifesting at birth with death from respiratory failure in infancy while the latter being diagnosed at 8-18 months of age or secondary to the delay of motor milestones in the face of normal intellectual development.

The presence of a second and nearly similar gene in the human genome (SMN2) ensures production of some SMN protein. Intellectual development and cognitive function of these children are not affected.

Alpha motor neurons are the most vulnerable to low SMN concentrations due to the high level of transcription that occurs, though other tissues may be affected as well.

SMA types 3 and 4 often manifest in older patients in whom there is deterioration of motor activity with muscle wasting and may mimic amyotrophic lateral sclerosis, though with the latter, there is muscular weakness that does not limit respiration nor eating nor cause wasting.

Congenital SMA with arthrogryposis is characterized by muscle weakness, floppy tone and contractures at birth and is invariably fatal.

Diagnosis of SMA is based on history: progressive motor weakness with profound muscle weakness is the hallmark of SMA. Resting muscle and tongue fasciculations are present in all forms of SMA and there may be a positive familial history of similar disease.

Genetic testing carried out by polymerase chain reaction (PCR) followed by restriction fragment length polymorphism method is the most accurate mode of diagnosis as it reveals total deletion of SMN1 in types 1 and 2 disease. Electromyography and muscle biopsy may be useful in ruling out other possible neuromuscular pathology.

The presence of SMN2 gene with its expression is thought to modify the effects of lack of SMN1 by increasing synthesis of the protein (SMN) that is neuroprotective and it forms the basis of novel drug and gene therapy interventions to ameliorate the effects of SMA. Currently, only intrathecal Nusinersen has been approved for clinical use.

Screening for SMA in utero and in neonates has also been carried out in a bid to plan early for gene therapy where necessary to enable expression of the SMN2 gene.

Anaesthetic Concerns

Airway: The airway of patients with SMA may be compromised due to the severe scoliosis limiting head flexion or neck extension as well as crowding of intraoral structures in the face of muscle wasting.

Respiratory system: Intercostal muscle wasting often means reduced vital capacity (restrictive lung disease) as well as inadequate ability to expectorate and clear secretions hence more frequent chest infections and a possibility of atelectasis postoperatively. Barotrauma may also occur if adherence to lung volume pressure ventilation curves is cursorily observed when mechanically ventilated.

Involvement of bulbar neurons may also compromise swallowing with resultant episodes of aspiration.

Cardiovascular system: As with many congenital anomalies, cardiac structural defects may occur though none has been noted to be specifically predominant in children with SMA.

However, the restrictive lung disease coupled with obstructive sleep apnea may predispose to pulmonary hypertension as well as a higher sensitivity among SMA1 and 2 patients to sedative drugs thus must have the same standard of care as general anaesthesia.

Neuromuscular System: These patients who have muscle atrophy due to neuronal deficiency undergo up regulation of nicotinic muscle receptors resulting in a tendency of developing hyperkalemia if muscle fasciculation occurs from the use of succinylcholine.

Involvement of bulbar motor neurons may also contribute to difficulty in swallowing with resultant malnutrition.

Due to reduced muscle mass, they tend to be very sensitive to the usual routinely used doses in clinical practice of neuromuscular non depolarizing agents and it is advisable to use the lower doses per kilogram body weight and where available, neuromuscular monitoring should be utilized. If possible and practical, the airway may be secured without the use of neuromuscular blockade.

The preferred agents would be the short acting steroids e.g. rocuronium or vecuronium (antidote is sugammadex) or alternatively, atracurium. Neuromuscular blockade must be reversed but bear in mind the possibility of a cholinergic crisis when neostigmine is utilized.

Positional challenges due to scoliosis and contractures may also occur affecting surgical site access or leading to a possibility of decubitus ulcers and/or pathological fractures associated with in cases of prolonged surgery.

Volatile inhaled anaesthetic agents have varying influence on skeletal muscle tone which is predominantly dose dependent relaxation and hence may temporarily worsen muscle control in SMA.

Total intravenous anaesthesia or target controlled infusion have the advantage of predictability especially when combined with multimodal analgesia.

The use of central neuraxial blockade may be both technically difficult due to the scoliosis, unpredictable distribution of anaesthetic agent as well as raise the possibility of further neuronal damage: peripheral nerve blocks have not been known to exacerbate SMA muscle weakness complaints and would be a good choice for analgesic management.

Postoperative pain: Opioids should be titrated and used with care if the pain scores deem it necessary in the face of adjunct analgesics.

Non-steroidal anti-inflammatory drugs, Paracetamol and regional peripheral anaesthetic blocks are the main stay of pain relief.

Thromboembolic phenomena in SMA due to inactivity have not been extensively studied and thus perioperative prophylaxis is individualized.

Conclusion

Spinal muscular atrophy type 2 disease is an autosomal recessive disorder which necessitates counselling of parents and early genetic testing to enable adequate supportive therapy. The supportive management of spinal muscular atrophy type 2 predisposes patients to multiple surgical interventions which pose a challenge in the face of central neuraxial blockade and the administration of neuromuscular junction blockade.

Postoperative analgesia should be multimodal with peripheral regional anaesthesia and non-opioids where feasible: opioids have been used but must be titrated for safety and closely monitored.

The difficult airway and ventilator mismatch are fairly common and must be anticipated when preparing for any sedative or anaesthetic exposure.

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06 Mar	Guiding Patient Therapy Through Clinical Diagnostics	Dr. Idris Chikophe Dr. Boitumelo Phiri	Thermofisher	Boma, Eldoret
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12 - 13 April	Simulation Training	Multiple	Abbvie	Kijabe Hospital
25 Apr	Insulins in Critical Care	Dr. Paul Ngugi	Eli Lilly	Double Tree
04 May	Medicolegal Symposium	Multiple	Several	Swiss Lenana Mount Hotel
16 May	Post-Operative Nausea and Vomiting		Sandoz	Panafric
06 Jun		TBC	Takeda	Panafric
15 Jun	Critical Care GAT Symposium	Multiple	Takeda	9 West
22 Jun	TCI/TIVA workshop		BD	
27 Jun	Sepsis		Sandoz	
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21 - 23 Aug	Think Global, Act Local: Changing Perspectives	Multiple	Multiple	Naivasha
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25 Oct	Post-Operative Nausea and Vomiting		Sandoz	
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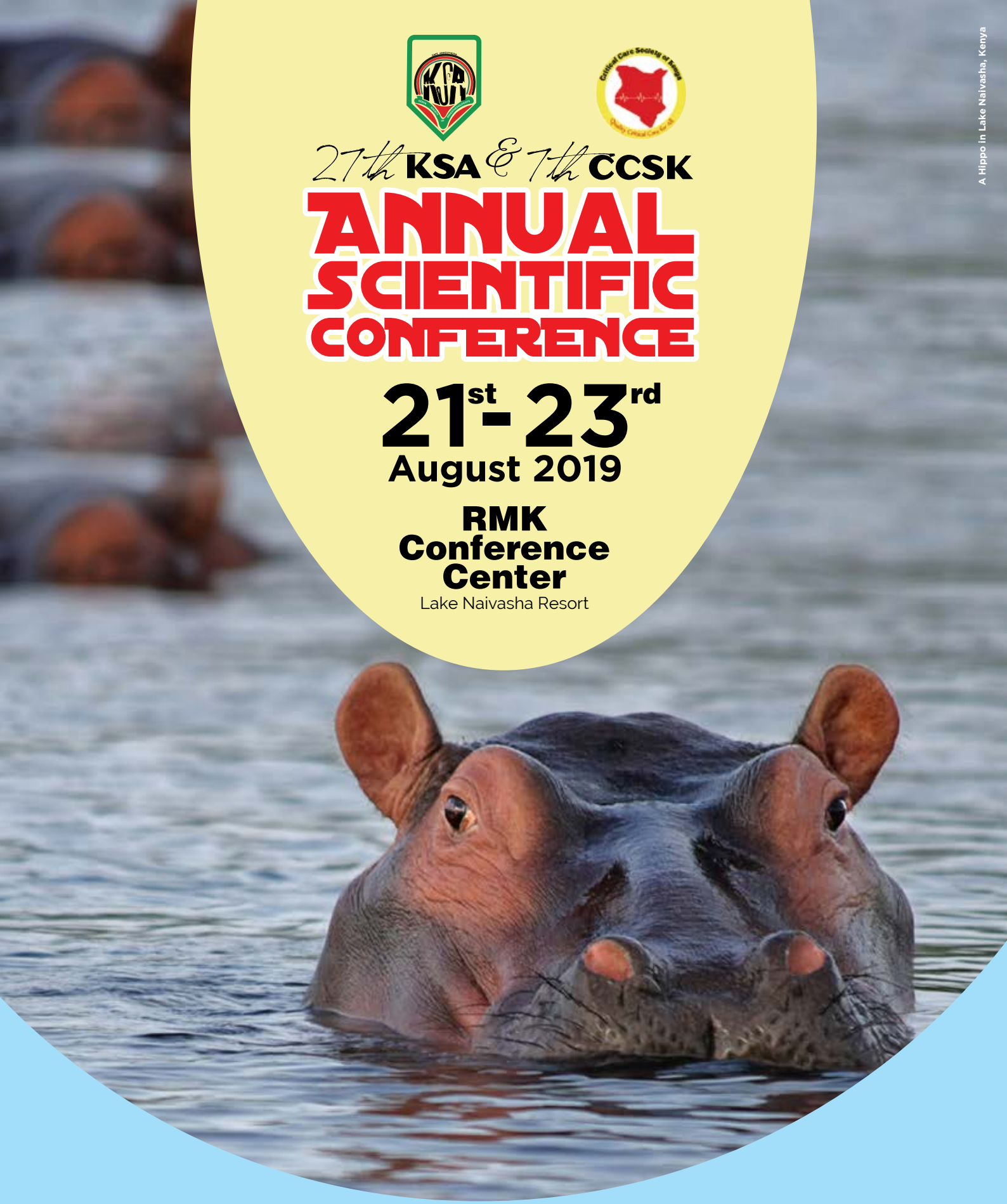

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