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There is no doubt that the Pharmaceutical Industry is doing a wonderful job, investing huge amounts of money in pursuit of new drugs and therapies to improve the quality of life and even the life span of our patients. But we, as physicians, must always be vigilant. The reason is that the main motive of many of the giants in the Pharmaceutical world is only profit. So, we come to this not uncommon scenario where a new drug gets licensed for sale for a particular indication. But once it is in the market a host of studies are undertaken to gather evidence to convince the licensing authorities and doctors that there are many more indications for use of the drug. The outcome is that near normal or subclinical states are classified as disease states and converted into targets for treatment. International Associations then jump on the bandwagon and bring out new guidelines suggesting that these subclinical or totally asymptomatic patients should be treated with these new agents. The number of people who subsequently get diagnosed or labelled as being “diseased” and requiring active treatment keeps on multiplying and the Pharma companies are laughing all the way to the bank.

We must be watchful against this trend of inventing diseases to sell drugs. Isn’t it time to pause and think carefully about the use of drugs with side-effects (not to mention costs and stress induced in the patient and family) in many of the recently created indications for therapy. Instead of concentrating only on laboratory investigations and ever changing targets for treatment, it is advisable to be a little old fashioned and assess the patient’s overall health and prognosis before rushing to treat adults and even children who are well, with medication. Remember the old saying, “do no harm” and reflect carefully on the indication for treating your patients – is it based on your clinical judgement and sound common sense, or is the decision inspired by data provided by Pharma sponsored studies and guidelines that encourage the use of drugs for subclinical states with doubtful benefit to the patient?
We are all lost in our own world and have no time or sympathy for other people. Often we are so immersed in our own problems that we become completely oblivious of the sufferings of others. Developing empathy and understanding other people’s problems and helping them in a meaningful way, will help enormously in calming our own mind and will go a long way in making the world a more peaceful place. To emphasize this point, Amma narrated the following story -

One day a man named Gopal who had a very good job in Dubai, suddenly lost his job due to the recession there. He was forced to return to his native place in Kerala. His wife and nine year old son also had to return with him. The whole family was very stressed at this sudden turn of events. Gopal began to apply afresh for jobs in Dubai. As the days went by he and his family became increasingly anxious. His wife would go every day to the nearby temple and pray fervently for Gopal to get another job, so that they could go back to Dubai and restart their lives as before. She promised the deity in the temple that if Gopal got another job then she would conduct a bhajan session in the temple.

After an extremely tense couple of months, Gopal got another job in Dubai. The relief and joy of his family is difficult to describe. Gopal’s wife kept her promise to the local deity and arranged a bhajan session at the temple.

Gopal, his wife and son, and other relatives all attended the bhajans. After they had sung seven or eight bhajans the session ended. But Gopal’s young son refused to leave. “Can we not sing at least two more bhajans?” he pleaded to his parents. “No, the session is over,” replied his father. His mother was also very firm. She said, “I had promised the deity that when your father gets another job, we will sing bhajans in the temple. Now he has got a job and we have finished singing and we have to go back home and pack our bags to return to Dubai.”

“But, mother,” pleaded the boy, “can we not sing just two more songs?” “Why are you so adamant?” asked Gopal.

The innocent boy replied, “Father, now you have got a job and our problems are solved, but can we not sing two more bhajans for so many other children whose father’s have also lost their jobs and who are suffering just as we had done for the last two months?”

Gopal and his wife were touched by their young son’s compassion for others. How many of us would spare a thought for others once our own problems are solved? It is this attitude of compassion that we need to have towards our patients. If we all had the attitude of the innocent boy then this world of ours itself will become heaven.
Norms for Crafting a Beautiful Smile

Priya K. Rahul D P, Varma S, Namitha R

ABSTRACT
A beautiful smile enhances self-esteem and also improves quality of life by providing overall health benefits. Smile can reflect the confident personality within an individual. A “Smile” decorates the esthetic aims of orthodontic treatment. Quest for an ideal smile has always an enigma for most orthodontists. This article has attempted to put forward different parameters for crafting a pleasant smile. It also tries to highlight different methods of smile analysis.

INTRODUCTION
Smile aesthetics has gained its importance with time. A smile is an outward sign of perceived self-confidence and internal satisfaction. Works by John Gottman has shown that smiling and other such expressions of positive emotions are important to shaping relationships with others.

In the past there have been different criteria for smile analysis and design. Early the clinical examination and diagnosis in treatment plan were largely focused on dental, and skeletal hard tissue element involved in a patient. The re-emergence of soft tissue paradigm in orthodontics has shifted diagnostic thinking to focus on soft tissue hard tissue interrelation and how they contribute to the overall facial aesthetic make-up of the patient.

DEVELOPMENT
The human smile, an inherited primate nature begins as reflex smile that develops without any stimuli within 1-3 days after birth. But a social smile that is in response to stimuli develops within 6-8 weeks after birth. Development of laughter is seen by 3 month after birth.

TYPE OF SMILE
Basically there are two types of smile Fig.1 that is 1) Duchene smile/Enjoyment smile-elicited by laughter or great pleasure, which results from maximal contraction of muscles causing full expansion of lips, with maximum anterior tooth display and gingival show, usually involuntary. 2) Posed smile/social smile-voluntary, unstrained, static facial expression, involving only moderate muscular contraction.

Another soft tissue determinant of the dynamic display zone is smile style. There are three smile styles -
1) Cuspid smile-characterized by action of all elevators of upper lip raising it like a window shade to expose the teeth and gingival scaffold.
2) Complex/full denture smile-that involves both upper and lower lip to display more teeth and gingiva, and
3) Mona-Lisa smile-by zygomaticus major muscle, drawing outer commissure outward and upward, followed by gradual elevation of the upper lip.

SMILE CAPTURE METHOD
Capturing patient smile images is a helpful diagnostic tool to study the patient’s lip-tooth relationship like smile arc, asses lip morphology with tonicity and other discrepancies during speech and smile. The ideal is that static (photographs) and moving (video) records be made. In the static records image gathering should include close-up shots in frontal, sagittal and oblique planes. Smile photographs are standardized in a natural head position, with eyes looking to a distant point, during the smile photograph. Both rest position and full smile photographs are taken.

Standardized digital videography allows the clinician to capture a patient’s speech, oral and pharyngeal function, and smile at the same time. By taking a video clip of both, we can evaluate all aspects of anterior tooth display.
Parameters for smile Analysis

Evaluating beauty is always subjective however; we need adequate tools to overcome the challenge of this subjectivity15. These criteria can be grouped into:

I) Facial Analysis
   A) Frontal view
   B) Profile view
II) Dento labial analysis
III) Dental Analysis
IV) Gingival Analysis

I) Facial analysis
Frontal view (Fig. 2)
   a) Relationship between interpupillary line and occlusal plane of teeth should be parallel.
   b) Relative position of soft tissue landmarks (nasal bridge, nasal tip, philtrum, chin point) and dental midline landmarks (upper incisor midline, lower incisor midline) should be on a line that is perpendicular to the frontal postural horizontal. Philtrum, usually least asymmetric of these points, is used as a starting point for midline structure assessment4.
   c) Lip symmetry (relationship of lips to face).
   d) Width of mouth—the ideal mouth is 50% of the width of face measured at mouth level4.
   e) Upper and lower lip lengths -The normal length from subnasale to upper lip inferior at relaxed position is 19-22 mm. The lower lip is measured from lower lower lip superior to soft tissue mention and normally measures in a range of 38 - 44 mm. The normal ratio of upper to lower lip is 1:2. Lip measurements identify normal or abnormal soft tissue length that can be related to dentoskeletal length normalcy, excess or deficiency13.

Profile view

a) Nasolabial angle - Angle is formed by the intersection of upper lip anterior and columella at subnasale. The normal angle should be at a desirable range of 850 to 1050. It helps to find estimation of lip tension present, antero-posterior lip thickness - normal lip thickness (10-11mm) & also skeletal discrepancies4.

b) Rickets E plane - It is drawn from tip of the nose to the chin. Ideally the upper lip and this plane should be at a distance of 4 mm and lower lip at a distance of 2 mm from this plane4.

c) Korkhaus Lip step—it is relation of upper lip to lower lip. Ideally upper lip is slightly ahead of lower lip. In class III malocclusion, there is protrusion of lower lip in relation to upper lip i.e. positive lip step. In class II malocclusion, negative lip step, with marked retrusion of the lower lip.

II) Dento facial Analysis (Fig.3)

a) Lip line - It is the amount of vertical tooth exposure in smiling – in other words, the height of the upper lip is relative to the maxillary central incisors5. “Tooth reveal” is a term for the amount of tooth structure or gingival that shows in various views and lip position. Even the most beautiful anterior tooth or teeth will have little esthetic value for the patient if the amount of reveal is unflattering to the face10. According to Morley youthful smiles reveal between 75% and 100% of maxillary central incisor crown proportion above the commissure line17.

Fig.3 shows A) tooth reveal B) gingival exposure, C) smile arc, D) buccal corridor and E) broadness of smile.

Ideal exposure with smile is three-quarters of the crown height to 2mm of gingival, females more than males. Variability in gingival exposure is related to 1) lip length, 2) vertical maxillary length and 3) magnitude of lip elevation with smile. Thus it can be average, high and low13.

In smiling, the upper lip is elevated by about 80% of its length, displaying 10mm of the maxillary incisor. Women have 3.5% more lip elevation than men. Thus care should be taken if a gingival smile is caused by a hyper mobile lip5.
b) Smile Arc-it is the relationship between a hypothetical curve drawn along the edges of maxillary anterior teeth and inner contour of the lower lip in posed smile, which is more pronounced for females than males. It can be classified as consonant (Fig.4), flat and non consonant /reverse – line. Care should be taken not to flatten smile arc during orthodontic procedure.

The patients whose lower lips touched or did not touch the incisal edges had a higher esthetic score than whose incisal edges were slightly covered5.

c) Smile symmetry - The relative positioning of the corners of the mouth in the vertical plane, can be assessed by the parallelism of the commissural and papillary line5. An esthetically pleasing smile usually shows symmetry and proportion between teeth, gingival and lips. The position of the mouth corners or lip commissures also affects the smile symmetry16.

The facial midline must coincide with the maxillary and mandibular central incisor midline or, minimally, these lines must be parallel. A small discrepancy of about 1.5 to 2 mm is acceptable, giving a natural appearance to dentition12. Also there should be no skewing of midline to right or left side which may be by transverse cant of the maxilla or a skeletal asymmetry.

d) Upper lip curvature - It is assessed from the central position to the corner of the mouth in smiling. Upward and straight lip curvatures are considered more esthetic than downward lip curvature4.

e) Buccal corridor/Negative space/vestibular space - Lateral negative space is the buccal corridor between the posterior teeth and the corner of the mouth in smiling5. The use of buccal corridor avoids the so called “16 teeth smile” or piano smile which characterizes a full mouth total prosthesis. This negative space is affected by the smile, the maxillary arch width, facial muscles, the position of the buccal surfaces of the posterior maxillary teeth, and also by the maxillary antero-posterior position related to the lips12. Smiles revealing upto the first molars were ranked the highest esthetically5. Archform also affects the transverse dimension of the smile. A broad arch is more likely to fill the buccal corridors than a narrow constricted arch.5

III) Dental Analysis

A pleasant smile along with its relationship between the teeth and lips also depends on the quality and beauty of the dental elements it contains and their harmonious integration. Dental components of smile include the size, shape, color, texture, alignment, crown angulations, midline, arch symmetry, contacts areas and embrasure5.

Tooth size is relative to face size and other teeth visual inspection and a rule of individual teeth being one sixteenth the dimensions of the face is a good starting point. When considering the width to height, the ideal maxillary central incisor should be approximately 80% with range of 66%-80%8. The dental arch is curved, less of each tooth is revealed toward the distal of the arch when viewed from frontal aspect. The ideal proportion of central incisor to lateral incisor to canine width should be 1.6:1:0.6. Using the Golden proportion, Lombardi ratio and Bolton analysis within the dental arch form can give guidance in where to add or subtract to create an esthetic look.

Subtle changes in the apparent size of teeth can be achieved by altering line angle, texture and color. Teeth can be made to appear thinner, longer, shorter, smaller or wider without changing the actual outline shape of tooth. The axial inclination of teeth becomes more medial as the teeth are further from the apparent midline and out into the buccal corridor and should be used together with correct axial alignment to produce a beautiful smile6.

The surface of teeth is textured or smooth. It determines light reflection and blending into other teeth. Placement of lines as developmental grooves or craze line and dimples can affect perception of width and length and alter light reflection pattern. Concave lines that run gingival to incisal increase perception of tooth height while lines that seen mesial/distal alter perception of tooth width. Line angles closes to the midline result in a shorter incisal edge, a small tooth face and larger embrasures, giving a small tooth. The maxillary incisors normally have mesial labial tilt and cuspids have pronounced lingual tilt with the gingival third appearing prominent6.

Arch position, shape, size and symmetry are also important. Square tapering arch of combines both square and tapering arch characteristic, with little crowding and overlapping of teeth. The incisor shows their full labial surfaces with cuspids having more distal rotation. Tapering arch is narrow from cusp to cusp with the
central incisor anteriorly placed. Ovoid arch resembles the tapering arch form but is wider from cuspid to cuspid forming an arc around the ridge. Since peg shaped/missing laterals are unaesthetic arc symmetry is also important.

Placement of contact areas is a critical aesthetic result in anterior teeth. Contacts (interdental contact points) are defined as the exact place that the teeth touch. Normal placement in maxillary anterior would be incisal third for the central incisor, incisal to middle third for central incisor to lateral incisors and the middle to gingival third for the lateral incisors to cuspids. The contact points progress apically as the teeth proceed from the midline to the posterior.

The connector (also referred to as the interdental contact area) is where the incisor and canine appear to touch. It tends to be 50% of the incisal-gingival length of the central incisor between the centrals, 40% between the centrals and the laterals and 30% between the lateral and canine. Embrasure form defines the outline of a tooth. The shape of embrasure alters the perception of tooth size such that large embrasures makes teeth look smaller embrasures make teeth look larger.

Even the shade and color patterns of the maxillary teeth follow a progressive pattern based on the distance from the midline. The maxillary central incisors are the lightest and brightest teeth in the smile. The maxillary laterals have a similar hue to that of the central incisors but are typically just slightly lower in color, or value. While canine have greatest chroma saturation and also are lower in value, premolars appear lighter and brighter than the canines, with value similar to that of lateral incisor.

### IV) Gingival Analysis

The gingival components of the smile are the colour, contour, texture and height of the gingiva. Dark colour gingiva is usually unaesthetics, the pink color gingiva is more esthetic. This problem is aggravated in gummy smile. Periodontal plastic surgery and gingival depigmentation is a solution for this. Gingival shape refers to curvature of the gingival margin of the tooth, determined by the cemento enamel junction and osseous crest. The gingival shape of the mandibular incisors and the maxillary laterals should exhibit a symmetrical half oval or half circular shape. The maxillary centrals and canines should exhibit a gingival shape that is more elliptical. Thus, the gingival zenith (the most apical point of the gingival tissue) is located distal to the longitudinal axis of the maxillary centrals and canine (Fig.5). The gingival zenith of maxillary laterals and mandibular incisors should coincide incisors should coincide with their lode with their longitudinal axis.

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**Diagram of Dental Aesthetic References - DDAR (new characteristics)**

DDAR (Fig.6) is an auxiliary diagnostic tool, that assist in the evaluation of mouth aesthetics, including reference of dental, gingival and labial structures, improving and facilitating the visualization of the smile. DDAR has intrinsically four horizontal lines, formed by the following structures-cervical line (gingival apexes), papillary line (papillary tips), contact point line (contact points), and incisal edge. The other two lines that make up the group of horizontal smile line are the upper lip line and lower lip line, thus composing the group of smile line.
1) Cervical line - The ideal form is attaining a convex aspect in relation to the occlusal plane. Because the apexes of the maxillary canines are most often higher than the lateral incisors and about the same level as the central incisors.

2) Incisal line - The incisal line follows the edges of anterior maxillary teeth. The ideal is that in young patients the incisal edges of the central incisors be below the edges of the lateral incisors and canines in a frontal view. In that configuration, the form of the incisal line resembles the outline of a “deep plate”. The classification of the incisal line includes concave (inverted plate), plain (shallow plate) and convex (deep plate).

3) Contact point line - The contact between anterior maxillary teeth is done in a descending fashion, starting from the canine. The line that unites these points will be almost parallel to the incisal line.

4) Papillary line - The papillary line is formed by the tips of the gingival papillae located between the canines and lateral incisors and between the maxillary lateral incisors and central incisors. It can be presumed that an ideal line would be parallel to the line formed by the contact points. The position of the papillae between the central and lateral incisor should be in an apical aspect in relation to that of the central incisors, as well as to the papilla of the lateral incisor and canine.

The best aesthetic relationship of anterior teeth is one that follows the 50-40-30 rule for the connecting space (Fig.7). As such, using the papillary line and contact points line as reference, we will have a band named “connector band”. The figure of this band resembles the shape of a “hang glide”.

5) Upper lip line - The ideal height of the smile line to be classified using as reference the relationship between the lower edge of the upper lip and gingival edge of the maxillary central incisor. However, 2 mm limit should be established above and below the gingival edge, thus instituting the three classes of smile height: high, medium and low.

6) Lower lip line - In general, it is the shape of the lower lip and the incisal edges of maxillary and mandibular teeth that create a pleasing or unpleasing smile ensemble. According to the parallelism between the arcs formed by incisal edges of anterior teeth and lower lip line, it can be consonant, flat and non-consonant. Also the vertical positioning of the maxillary incisors and canines forms a curvature; the line that contours this relationship resembles a deep plate.

Several other factors mentioned earlier also needs to be taken in consideration for evaluating a esthetic smile.

Thus smile, though a simple act involving mainly zygomatic major, involves the evaluation of certain elements that aids a dentist to diagnose and create a good esthetic smile. Recent advances in technology permits the clinician to measure dynamic lip - tooth relationships and incorporate that information into orthodontic problem list and biomechanical plan. Successful smile designing requires an understanding of the patient’s soft-tissue treatment limitations and the extent to which orthodontics or multidisciplinary treatment can satisfy the patient’s and orthodontist’s esthetic goals.

CONCLUSION

As public awareness of esthetic dental treatment increases patient seek to enhance and resolve several common concerns to achieve a good smile. Orthodontic case in which occlusion meets every criterion of American Board of Orthodontics for a successful treatment may not produce an esthetic smile. Thus the goal of a clinician is to create not only an admired look, but also the ability to harmonize with hard and soft tissues.

Patient’s concern and opinions on esthetic have to be given prior importance. On this matter Wylie astutely wrote that “the layman’s opinion of the human profile is every bit as good as the orthodontist’s and perhaps even better, since it is not conditioned by orthodontic propaganda”. Recent advances like a smile evaluation helps in this concern, which include the parameters of a smile analysis that help to create a beautiful smile along with considering patient’s perception on esthetics. Smile variation among different races is a subject of future research.
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Anaesthesia care beyond Operating Rooms: Newer opportunities & Challenges.

Ramkumar P

Technological progress in medical science leads to more interventional procedures, with which we are able to give symptom relief for sicker patients who may not be a candidate for a major surgical procedure. The risk involved in providing anaesthesia for the sicker patients for the interventional procedure is much more than the regular surgery. As a critical care and resuscitation expert and with special skills/ knowledge of Anaesthesia, the services of the anaesthesiologists are demanded by non surgical specialists who do not make use of the regular operating rooms for the work.

Providing anaesthesia care in out of operating room situations may be challenging, as changed and variable environments pose unique problems. When providing care at such locations, anesthesiologists must maintain the same high standard of anesthetic care provided in the operating suite. The anaesthetizing location must be surveyed by the anaesthesiologist to determine whether anaesthesia care can be delivered safely in that location before delivery of that care. The requirements for anaesthesia and the patient’s underlying condition do not vary merely because of location, but the conditions under which the anaesthesia care is delivered may vary greatly because of the space and equipment available in these locations. Large, mobile pieces of radiologic equipment, radiation hazards, intense magnetic fields, paramedical personnel not familiar with the anaesthesia team, and other factors may compromise the delivery of quality anaesthesia care.

Despite the availability of detailed guidelines, a recent analysis of closed anaesthesia claims demonstrated greater injury severity and more substandard care than seen with operating room closed claims. Drug interactions were the most common associated factor, followed by drug overdose, inadequate monitoring, inadequate skills for cardiopulmonary resuscitation, inadequate evaluation before sedation and premature discharge from medical supervision were other incidents noted as contributory to the events in outside or procedures. Presence of a dedicated person for monitoring the patient and following the proper guidelines can minimize the events.

Guidelines for anaesthetic care delivered outside the Operating Room American Society of Anaesthesiologists (ASA) 1994 Guidelines for Non–Operating Room Anaesthetizing Locations include recommendations for

1. A reliable oxygen source with backup as required.
2. A working suction source with all proper connections & suction catheters.
3. Waste gas scavenging system.
4. Adequate monitoring equipment to meet the standards for basic anaesthesia monitoring and, in addition, a self-inflating hand resuscitator bag/ transport ventilator.
5. Sufficient safe electrical outlets.
6. Adequate illumination of the patient and anaesthesia machine with battery-powered backup.
7. Sufficient space for the anaesthesia care team.
8. An emergency cart with a defibrillator, emergency drugs, and other emergency equipment.
9. A means of reliable two-way communication to request assistance.
10. Compliance of the facility with all applicable safety and building codes.

It is the responsibility of the anaesthesiologist providing care to ensure that the anaesthetizing location in which that care is delivered meets all applicable standards.

Patient population:
Wide range of patients from newborn to geriatric patients, healthy to critically sick patients will be subjected for the anaesthesia care outside or procedures. Children, unconscious / uncooperative or anxious patients, elderly or confused patients all require anaesthesia care. The reasons may be needle phobia, claustrophobia, painful procedures or procedures requiring absolute immobile patient, comorbidity requiring constant monitoring and resuscitation during the procedure. When very advanced and costly procedures are being carried out physicians and patients prefer TOTAL CARE especially when the additional expenditure for anaesthesia care is negligible compared to the procedure charges.

Monitoring:
ASA standards for basic anaesthesia monitoring require presence of qualified anesthesia personnel throughout conduct of the course of anaesthesia and continuous evaluation of the patient’s oxygenation, ventilation, circulation, and temperature. Provision is made for the absence of
anaesthesia personnel from the immediate vicinity of the patient if required for safety (i.e., in the presence of radiation hazards), provided that adequate patient monitoring is continued despite the physical separation of the anesthesiologist from the patient. Oxygen concentrations of inspired gas should be monitored with the use of a low-concentration alarm, blood oxygenation should be monitored with pulse oximetry, and ventilation should be monitored by observation and by detection of end-tidal carbon dioxide. Continuous end-tidal carbon dioxide analysis should be performed. When mechanical ventilation is used, a disconnect alarm with an audible signal must be present. Circulation is monitored by continuous display of the electrocardiogram, as well as by measurement of arterial blood pressure at a minimal interval of 5 minutes, in addition to other assessments such as auscultation, palpation of pulse, invasive blood pressure monitoring, or oximetry. When changes in body temperature are anticipated or suspected, patient temperature should be assessed. There should be no hesitation to use invasive monitoring if the patient condition warrants so in case for or procedure.

GOALS:
The goals of sedation/anaesthesia outside or can be summarized as follows

- Guard the patient’s safety and welfare
- Minimize Physical discomfort and pain
- Control anxiety, minimize psychological trauma and maximize the potential for amnesia
- Control behavior and/or movement to allow safe completion of the procedure
- Return the patient to a state in which safe discharge from medical supervision is possible.

Problems:
Unfamiliar locations and working conditions pose certain problems like

- Related to physical layout of the facility
- Remoteness from available help.
- Difficult or limited access to patients.
- Unfamiliar or outdated anaesthesia equipment.
- Untrained personnel.

General Precautions:

- Proper check up of anaesthesia machine & equipment
- Availability of adequate number of gas cylinders
- Obsolete and poorly functioning equipment should be discarded.
- Proper grounding of electrical equipment
- Availability of adequate persons and materials for the procedure and monitoring

- Facility for post procedure care/PACU.

Patient Evaluation:
Clinicians should be familiar with the sedation-related aspects of the patient’s medical history.

These include (1) abnormalities of major organ systems, (2) previous adverse effects with sedation and general anesthesia, (3) drug allergies, current medications, and drug interactions, (4) time and nature of oral intake, and (5) history of tobacco, alcohol, or substance abuse.

A focused physical examination including vital signs, auscultation of the heart and lungs, and evaluation of the airway is recommended.

Preprocedural Preparation:
Patients should be informed of and agree to sedation, including its risks, benefits, limitations, and alternatives. Sufficient time should elapse before a procedure to allow gastric emptying in elective patients. Minimum fasting periods of 2 hours (clear liquids), 4 hours (breast milk), and 6 hours (infant formula, nonhuman milk, and light meal), are recommended for healthy patients. If urgent, emergent, or other situations impair gastric emptying, the potential for pulmonary aspiration of gastric contents must be considered in determining the target level of sedation, delay, or intubation.

Medications: Monitored anaesthesia care (MAC), general anaesthesia (GA) or regional anaesthesia may be required. Midazolam, Fentanyl, Propofol, and Ketamine are frequently used drugs. Dexametomidine is useful for conscious sedation as well as for facilitating smooth anaesthesia and recovery when GA is needed.

A moderately sedated child who can respond to light touch can protect his or her airway and a deeply sedated child who can respond appropriately only to pain may not be able to control the airway. The important assessment of the child is not response to stimulation but the ability to protect the airway. Different sedative drugs have differing effects on analgesia versus airway obtundation. Propofol is not a profound analgesic but has profound effects on the airway. Conversely, the sedative drug Dexametomidine may provide profound sedation with little depression of respiratory function. Ketamine produces intense analgesia and most children maintain a patent airway and adequate respiratory effort.

When Spinal anaesthesia is needed for day care patients, ropivacaine is a better choice due to less motor block and early recovery.

Combinations of Sedative/Analgesic Agents: Combinations of sedative and analgesic agents may be administered as appropriate for the procedure being performed and the condition of the patient. Ideally, each component should be administered individually
to achieve the desired effect (e.g., additional analgesic medication to relieve pain; additional sedative medication to decrease awareness or anxiety). The propensity for combinations of sedative and analgesic agents to cause respiratory depression and airway obstruction emphasizes the need to appropriately reduce the dose of each component as well as the need to continually monitor respiratory function.

Titration of Intravenous Sedative/Analgesic Medications: Intravenous sedative/analgesic drugs should be given in small, incremental doses that are titrated to the desired end points of analgesia and sedation. Sufficient time must elapse between doses to allow the effect of each dose to be assessed before subsequent drug administration. When drugs are administered by nonintravenous routes (e.g., oral, rectal, intramuscular, transmucosal), allowance should be made for the time required for drug absorption before supplementation is considered. Because absorption may be unpredictable, administration of repeat doses of oral medications to supplement sedation/analgesia is not recommended.

Anesthetic Induction Agents Used for Sedation/Analgesia (Propofol, Methohexital, Ketamine): Even if moderate sedation is intended, patients receiving propofol or methohexital by any route should receive care consistent with that required for deep sedation. Accordingly, practitioners administering these drugs should be qualified to rescue patients from any level of sedation, including general anesthesia. Patients receiving ketamine should be cared for in a manner consistent with the level of sedation that is achieved.

Anxiolytics/Sedatives

The most commonly used anxiolytics/sedatives in pediatric sedation are chloral hydrate, diazepam, and midazolam. Chloral hydrate is one of the most widely used sedatives in neonates and children younger than 3 years of age. It is widely used as a sedative to facilitate nonpainful diagnostic procedures such as EEG and CT or MRI. It is rapidly and completely absorbed when given orally. Rectal administration is erratically absorbed and therefore not recommended. Onset of sedation is 30 to 60 minutes, and the usual clinical duration is 1 hour. Although it has a long safety record, it can cause respiratory depression due to airway obstruction, and deaths have been associated with its use alone and when combined with other sedating medications.

The benzodiazepines are commonly used in pediatric sedation. They are anxiolytic, amnestic, sedative hypnotics with anticonvulsant activities but no analgesic properties. Their high lipid solubility at physiologic pH accounts for the rapid CNS effects. As opposed to diazepam, midazolam is delivered in a water-soluble form (pH 3.5), which markedly decreases the incidence of pain on injection and thrombophlebitis. However, the resulting decrease in fat solubility markedly delays transport into the CNS (peak EEG effect 4.8 minutes for midazolam versus 1.6 minutes for diazepam). The sedated child usually becomes compliant but does not lose consciousness. Children frequently move, and another agent, such as an opioid, may be necessary if the child must not move to successfully accomplish the procedure. The benzodiazepines have the advantage of antegrade amnesia in a significant number of patients. The markedly prolonged and variable elimination half-life and active metabolite of diazepam (desmethyl-diazepam) make midazolam a superior sedative drug in children, particularly infants. Time to peak effect after intravenous administration of midazolam is 2 to 4 minutes, and duration is 45 to 60 minutes. Midazolam can be given intravenously, intranasally, orally, or rectally. It is the only drug in this class approved for neonates. Benzodiazepines produce mild respiratory depression and upper airway obstruction. Respiratory depression may become severe in compromised children or in children with tonsil hypertrophy. Benzodiazepines must be given after appropriate guidelines. The combination of benzodiazepines and opioids is particularly troubling because they can produce a “super additive effect” on respiratory depression.

Flumazenil is a specific benzodiazepine receptor antagonist and will rapidly reverse the sedative and respiratory effects of benzodiazepines. It is the first specific reversal agent for benzodi-azeines and rapidly reverses CNS-induced unconsciousness, respiratory depression, sedation, amnesia, and psychomotor dysfunction. The recommended dose of flumazenil is 10 μg/kg up to 0.2 mg every minute to a maximum cumulative dose of 1 mg intravenously. Antagonism begins within 1 to 2 minutes and lasts approximately 1 hour. Because re sedation after 1 hour may occur, the child must be carefully monitored for at least 2 hours. Repeat flumazenil may be necessary. It should be noted that flumazenil will not antagonize respiratory depression due to opioids. Flumazenil should not be administered for the routine reversal of the sedative effects of benzodiazepines but reserved for reversal of respiratory depression.

Barbiturates:

Pentobarbital is the most commonly used intermediate-acting barbiturate for sedation. It has no analgesic effect and produces sedation, hypnosis, and amnesia. It has a long history of use during radiologic procedures. Sedation starts in 3 to 5 minutes and peaks in 10 minutes. Studies have shown a low incidence of respiratory obstruction and transient desaturation as well as hypotension. The barbiturates tend to make children more sensitive to pain and should be combined with analgesics when used during painful procedures. Newer, shorter-acting, faster recovery drugs are quickly replacing pentobarbital.
Opioids:

Opioid analgesics are rarely used alone for diagnostic and therapeutic procedures in children. These potent analgesics are important during painful diagnostic and therapeutic procedures. They bind with four primary opioid receptor types (mu, kappa, delta, and sigma) that are located in the brain, spinal cord, and periphery. Serious effects of opioids include respiratory depression, bradycardia, hypotension, seizures, and opioid-induced glottic/ chest wall rigidity.

Morphine may be considered for painful procedures (>1hour) or when the child will also be in pain after the procedure. The duration of action is 3 to 5 hours after intravenous administration. Morphine may be given orally (0.2 to 0.5 mg/kg), intravenously (0.05 to 0.1 mg/kg [maximum 0.3 mg/kg]), or intramuscularly (0.1 to 0.2 mg/kg). Time to peak effect for oral, intravenous, or intramuscular administration is 60 minutes, 3 to 5 minutes, and 10 to 30 minutes, respectively. Its slow onset and prolonged duration have caused it to be replaced by shorter acting opioids when used for sedation and analgesia for procedures.

Fentanyl has replaced morphine as the opioid of choice for analgesia/sedation for procedures in children. Intravenous fentanyl is a potent pure opioid (i.e., 100 times more potent than morphine) with no amnesic properties. Its high lipid solubility allows for onset within 30 seconds and a peak effect at 2 to 3 minutes. It has a brief clinical duration of 20 to 40 minutes when given in small doses owing to its rapid redistribution to skeletal muscle, fat, and other inactive sites. Unlike morphine it has no active metabolites. Fentanyl’s clearance is decreased and its half-life is increased in preterm and term infants. It is fully reversed by opioid antagonists and is frequently used with a short-acting anxiolytic (midazolam). Intravenous doses usually start at 0.5 to 1 μg/kg and are titrated every 5 minutes to effect but not to exceed 5 μg/kg. Doses must be given in small aliquots and carefully titrated to avoid chest wall and glottic rigidity. Close post-procedural observation is required because respiratory depression can outlast analgesia.

Remifentanil is the newest rapid-acting opioid. This rapid onset, extremely potent, lipophilic short-duration opioid is metabolized by plasma cholinesterase. Remifentanil has been used for intraoperative sedation by anesthesiologists and in intubated children in the ICU. Remifentanil is associated with a high incidence of apnea and chest wall rigidity and should not be used by the non-anesthesiologist for pediatric sedation.

Opioid antagonists specifically reverse the respiratory and analgesic effects of opioids and should be readily available when opioids are used. Naloxone is the most commonly used an-agonist. Opioid antagonists should not be used for routine reversal of the sedative effects of opioids but reserved for reversal of respiratory depression / respiratory arrest. Naloxone may be given intravenously, intramuscularly, or subcutaneously. The initial dose for respiratory depression is 0.01 mg/kg titrated to effect every 2 to 3 minutes. Ten to 100 μg/kg up to 2 mg may be required for reversing respiratory arrest. Adverse reactions from reversal of analgesia include nausea, vomiting, tachycardia, hypertension, delirium, and pulmonary edema. Patients on long-term opioid therapy should be given opioid reversal agents in low doses and with extreme caution because withdrawal seizures and delirium may occur. If naloxone is used, then the patient should be observed for a minimum of 2 hours. Repeat naloxone may be necessary. Nalmefene (Revex) has a longer half-life (~10 hours) than naloxone. Its half life outlasts the effects of fentanyl and negates the treatment of pain with opioids for several hours.

Systemic Anesthetics

These drugs should be used only by anesthesiologists or other practitioners who have specific training in their use and have advanced airway management skills because airway obstruction, apnea, and cardiovascular instability may quickly and unpredictably occur. Ketamine is one of the few sedatives that produce both amnesia and analgesia. The clinical appearance is that of a patient who has opened eyes (usually with horizontal nystagmus) but does not respond to pain. Ketamine has been shown to preserve cardiovascular function in most cases and to have limited effects on respiratory mechanics and allows for spontaneous respirations. Ketamine is associated with nonpurposeful motion, which limits its usefulness when immobility is necessary (e.g., use during CT). Ketamine can markedly increase cerebral blood flow and is contraindicated in patients with increased intracranial pressure. Other contraindications include those with head injury, open globe injury, hypertension, and psychosis. Ketamine can decrease the response to hypercarbia, as well as cause laryngospasm, coughing, and apnea. No antagonist is available. Typical starting doses are 1 to 2 mg/kg intramuscularly, 0.25 to 1.0 mg/kg intravenously, or 4 to 6 mg/kg orally. The onset after intramuscular injection is 2 to 5 minutes, with a peak of 20 minutes; duration can be 30 to 120 minutes. Onset after intravenous administration occurs in less than 1 minute, with a peak effect in several minutes and duration of action of approximately 15 minutes. Oral doses of 4 to 6 mg/kg are usually combined with atropine and have an effect in 30 minutes and last up to 120 minutes. Larger doses or supplementation with other sedatives or opioids may produce deep sedation/general anesthesia. Ketamine should be administered with an antisympathomimetic (atropine, 0.02 mg/kg, or glycopyrrolate, 0.01 mg/kg) because copious secretions from ketamine alone may induce laryngospasm. Although initially thought to maintain airway reflexes, this is not always
the case; ketamine may not protect against aspiration. Unpleasant dysphoric reactions (so-called emergence reactions) (up to 12%) can be severe but are usually mild. The prophylactic use of midazolam does not decrease this incidence.

Etomidate produces sedation/anesthesia, anxiolysis, and amnesia similar to the barbiturates and propofol. Its major advantage is its lack of adverse cardiovascular effects. Loss of consciousness occurs in 15 to 20 seconds, and recovery is due to redistribution and occurs in 5 to 10 minutes. Etomidate has been used in adults and children for procedural sedation, although the end point of sedation is not well described and often is general anesthesia. Transient adrenal suppression can occur after multiple doses and after single dose administration. Propofol is an anesthetic that is widely used for pediatric sedation and anesthesia. Its onset is within 30 seconds. It is highly lipid soluble and the lipid solubility makes the drug effect diminish extremely quickly (5-15 minutes). It has no analgesic properties, but it does have antiemetic and antipruritic properties. Although small doses of propofol (25-50 μg/kg/min) can provide moderate sedation in adults, deep sedation and airway obstruction quickly occur in children. Dosing in adults for sedation is recommended at 25 to 200 μg/kg/min, whereas many children require considerably higher doses. Propofol is a profound respiratory depressant and can lead to rapid airway obstruction and apnea. Other adverse reactions include increased salivary and tracheobronchial secretions, myoclonic movements, anaphylactic reactions, and bacterial contamination. Pain on injection can be lessened by the addition of lidocaine to the solution. Hypotension is mild and usually not clinically significant in normal healthy patients. Durations of propofol for more than 5 hours have been associated with propofol infusion syndrome but not during shorter procedures for sedation. Patients should be assumed to be deeply sedated or anesthetized when using this drug, and it should be administered only by practitioners with advanced airway skills. Propofol “procedural sedation” delivered by nonanesthesiologists is growing rapidly in intensive care units, emergency departments, dentistry, oral surgery, and gastroenterology suites. The dosing recommendations for use in “procedural sedation” in these areas frequently do not cause sedation but rather anesthesia. (“Procedural sedation” is defined as a depressed level of consciousness but one that allows the patient to maintain airway control “independently and continuously.”) However, multiple studies show that propofol procedural sedation frequently causes anesthesia with inability to maintain the airway. The controversy over whom and under what conditions propofol (or other potent sedatives) should be administered for procedural sedation needs to be addressed and agreed upon on a national level.

Nitrous oxide (N2O) is a potent inhalation analgesic with a peak effect in 3 to 5 minutes and very rapid return to baseline when discontinued. A premixed tank of no more than 50% N2O is available (Entonox). Administration of N2O can be used for “minimal sedation”. (1) only ASA-PS I or II patients; (2) only 50% nitrous oxide or less is used; (3) inhalation equipment must have the capacity to deliver 100% oxygen and never less than 25% oxygen; and (4) a calibrated oxygen analyzer must be used. Although N2O in 50% concentration with oxygen usually produces “minimal” sedation, the addition of any sedatives/hypnotics may rapidly produce a deeper level of sedation and require increased monitoring and vigilance.

Dexmedetomidine is an imidazole 02 agonist that is similar to clonidine but with an even higher 02 : 01 specificity ratio of 1600 : 1. Its elimination half-life in children is 2 hours. The drug is highly lipid soluble and quickly crosses the blood-brain barrier. Its CNS effect is to stimulate receptors in the medullovasomotor center, which decreases sympathetic tone. It also stimulates central parasympathetic outflow and decreases sympathetic outflow from the locus ceruleus of the brainstem. The decreased outflow from the locus ceruleus allows for increased activity of the inhibitory GABA neurons, which cause sedation and analgesia. Dexmedetomidine is approved for sedation of ventilated adult patients in the ICU but not in children. When administered in clinical doses it causes limited effects on ventilation in adults and may mimic natural rapid-eye-movement sleep. The initial dose must be given over 10 minutes, followed by an infusion. When given in the recommended fashion it decreases blood pressure and heart rate in adults. It should be used with caution in children with preexisting bradycardia, atrioventricular conduction defects, hypotension, and decreased cardiac output. Dexmedetomidine may provide safe sedation for procedures with minimal effect of the airway and therefore markedly improve safety.

Patient transport to Recovery Room/ post anaesthesia care unit:

The patient must be medically stable before transport. The patient must be accompanied to the recovery area by the individual providing the anesthesia or sedation/analgesia care, and monitoring used according to the patient’s medical condition must be maintained. Provision of oxygen delivery and monitoring while the patient is on the transport cart may be required. Appropriate recovery facilities and staff must be provided. In the recovery area, the patient’s condition must be documented and continually assessed. Immediate availability of personnel trained in advanced cardiac life support should be ensured. Patients should not be discharged until they have met specific discharge criteria. Clear directions are to be given in writing and explained to the care taker at the time of discharge.
1. Anaesthesia for diagnostic neuroradiological procedures:

- Computed Tomography (CT)
- Magnetic Resonance Imaging (MRI)
- Pneumoencephalography
- Angiography: Includes mainly spinal cord and cerebral angiography

Patients in the radiology suite may have severe underlying medical conditions such as cardio-vascular, pulmonary, or neurologic disease. Indeed, they may be in the radiology suite, as opposed to the operating suite, precisely because their severe underlying disease precludes operative intervention. Finally, anesthesiologists may be summoned relatively late in the care, after failure of sedation/analgesia administered by the radiologist or nonanaesthesia personnel. Clearly, this situation is undesirable, and open communication between the departments of radiology and anesthesiology is essential. Special precautions should be taken in MRI due to the effect of magnetic field on ferrous objects.

2. Anaesthesia for interventional radiology.

Angiograms, angioplasty and angio-embolization are becoming a regular work in many radiology labs. Most of the procedures can be done as a Monitored Anaesthesia Care and moderate sedation / Conscious Sedation, but some require complete immobilization and require General Anaesthesia with controlled ventilation. Radio Frequency Ablation of the solid tumour mass in the lungs, Liver and bones is another area where similarly anaesthesia service could be utilized. Some of the RFA patients will benefit from continuous regional block with catheter for Post procedural pain relief. The iodinated contrast media used in the radiology and neuroradiology suites, as well as the cardiac catheterization laboratory, may cause significant adverse reactions, and patients receiving contrast media require close monitoring.

3. Anaesthesia for radiotherapy

Intra Operative Radiation Therapy (IORT) & external beam radiation; MAC or moderate to deep sedation may be needed for anxious patients to keep without moving during the CT simulation and External RT especially in paediatric patients who may require 5/7 days sedation/ TIVA that too for a month or so depends upon the number of fractions needed. Peripherally inserted CV Access will be useful in these patients.

Intra Cavitary Radio Therapy (ICRT/ BRACHY- THERAPY): Compared to Low Dose Rate therapy taking nearly 24 hours time the High Dose Rate with Iridium 192 the treatment time is only 20 minutes and the whole process will be over in 3-4 hours time. ICRT will be used in Carcinoma Cervix patients and these patients could be managed as Day Care patients under regional anaesthesia /CS/GA. Subarachnoid Block with ROPIVACAINE and FENTANYL works for the period of complete treatment and avoids involuntary movement of the lower limbs causing displacement of the applicator / implants and if there is any misplacement it allows to correct the applicators after confirming under IMAGE.

4. Coronary angiography and cardiac catheterization,

5. Anaesthesia for electroconvulsive therapy (ECT),

6. Anaesthesia in Emergency Room/ Trauma care Unit,

7. Anaesthesia in Emergency Room/ Trauma care Unit,

8. Anaesthesia for IVF & GIFT: To be aware of the Ovarian Hyper Stimulation Syndrome.


10. Anaesthesia for children in Oncology/ Rheumatology/ Nuclear Medicine etc.

Anesthesia care in remote locations: These include war fronts and other mass casualties, including disasters and terrorist violence etc. The guiding principles of care are same as envisaged in “minimum monitoring and safety standards”, advocated by the ISA. There is no justification in giving anaesthesia (other than field and local blocks) without ensuring the availability of a reliable oxygen source, facilities to establish definitive airway and pulse oximetry. On the other hand resuscitation for basic and advanced life support and comprehensive trauma life support (CTLS) should be undertaken whenever and wherever necessary and feasible. The techniques and extent of the life support instituted will depend upon the place, available facilities and resuscitator’s expertise.

Conclusion: The role of anaesthesia outside the operating rooms is rapidly expanding and evolving along with the advances in interventional radiology and other invasive modalities. However, we must understand that there are many constraints, as the co-morbid conditions of the patients are similar and often more severe than what we face in the operating rooms, and in-creasingly complex diagnostic and therapeutic procedures are being performed on sicker patients. Understanding the anaesthetic constraints and complexities and keeping abreast with the current developments are crucial in ensuring the maximal benefits to and safety of the patients.

Suggested reading:

Miller’s Anesthesia Seventh edition: Ronald D Miller et al, Chapter 79 – Anesthesia at Remote Locations, Paul E.Stensrud

Voice Clinic Utility and Performance: Institutional experience

M. P. Cherian, U. K. Menon

ABSTRACT

Background: Patients with voice complaints can benefit from dedicated care given in a voice clinic.
Aim: To assess the usefulness of the voice clinic in the ENT OPD and Audiology Dept. at Amrita Institute of Medical Sciences
Methods: Retrospective collection of data
Conclusion: Satisfactory audit of the subjective and objective success of patients managed in the voice clinic

INTRODUCTION

Voice forms part of human identity. Any problem with it results in multi-dimensional handicap for the patient. Prevention, thorough analysis, investigation and relevant correction are all needed to address this problem. In the present-day health care scenario, a dedicated Voice Clinic has become an undoubted necessity in an Institution such as A.I.M.S.

Relevance of the condition:

Increased awareness of the concept of quality-of-life has resulted in the hitherto ignored symptom of change of voice presenting much more commonly in the ENT OPD. The various patient groups seen are:

- Teachers, with varying degrees of vocal strain
- Voice professionals, including TV presenters, lawyers
- Singers
- Non-voice professionals with hoarseness
- Children with voice abuse issues

The importance given by the patient and his/her social environment to the change in voice is considerable. Besides, it also raises issues of professional competence for the affected person. Needless to say, resolution of this complaint is a ‘felt need’, and a challenge to all the health professionals involved. This is where a focused team in a Voice Clinic has its role.

Incidence:

Exact figures are not available in the Indian context, but roughly 10% of the general population experience some sort of voice problem at one time or the other. Burden of the problem in our ENT OPD is proportionately higher. A dedicated Voice Clinic helps to further augment the flow of such patients here.

Management modalities:

A] At the average ENT facility
- Laryngeal examination [mirror and/or endoscope]
- Referral for voice therapy to an Audiology Speech Therapy centre

B] At a Hospital with ENT Department
- Rigid and/or flexible laryngoscopy with a record [images or video] given to patient
- In-house Speech Therapy management
- Surgery, where indicated
- Referral to higher centre for complicated cases

C] At a higher/tertiary referral centre
- Flexible laryngoscopy + possibly, laryngostroboscopy
- Voice analysis in the Audiology Department
- Microlaryngeal and phonosurgery

D] In a Voice Clinic

All of the above, followed by:
- Video Laryngostroboscopy (VLS)
- Detailed voice analysis using specific software
- Relevant consultations to other Departments [Pulmonology, Neurology]
- Patient seen in the Clinic by both Laryngologist and Speech Therapist
- Focused attention on the patient and his/her voice issues
- Detailed discussion and voice therapy advice
- Regular follow-up in the Clinic

Present situation in AIMS:

A rudimentary voice clinic was started by late 2009. With the procurement of a Stroboscopy machine (Atmos) and an internationally used software programme for Voice analysis (Dr. Speech), the Clinic became fully operational by mid-2010.

Present protocol:

i) New patient with a voice complaint seen in the routine ENT OPD.

Voice-specific details entered by the encountering Doctor in a Voice Proforma, which has been specifically added to our AHIS. In case a rigid or flexible laryngoscopy is done, the video is recorded for later reference.
ii) Patient given appointment to be seen in the Voice Clinic.
iii) In the ENT OPD, thorough evaluation including a Videolaryngostroboscopy (VLS), which is the cornerstone of a Voice Clinic, is performed.
iv) A subjective questionnaire, the voice handicap index (VHI-30), is given to the patient. This is an internationally accepted form consisting of 30 questions, related to the subjective difficulty experienced by the patient, due to his/her voice problem. We are using a translated Malayalam version of the questionnaire. A validation study has been done for the same.
v) Voice analysis includes recording of the voice using Dr. Speech. In certain cases, especially for speech, a free software programme called Praat is used. These are done in our Audiology Dept. The former software is also used in treatment programmes.

Treatment:

**Counseling:** This is the first step which is done in the clinic. The patient is made aware of the basics of the functioning of the voice box, and the routine steps to maintain a healthy voice and prevent voice disorders.

**Voice therapy:** This is the cornerstone of treatment of most voice problems. Our specially trained Voice therapy team, in the Audiology Dept., administers specific techniques to be practiced by the patient at home, and at follow-up sessions. Majority of the conditions, like vocal nodules, voice strain, muscle tension dysphonia etc. resolve fully with VT.

**Surgery:** Certain cases would need surgical correction. These could range from benign organic lesions on the vocal folds to structural and neurological problems. The entire spectrum of surgeries is given the umbrella heading of ‘Phonosurgery’.

Relevance of the special investigations in Voice Clinic:

**A] Stroboscopy**

The ‘strobe effect’ is the apparent slowing down of a rotating or vibrating body when flashing light of approximately the same frequency falls on it. It is an optical illusion, which is made use of to look at the moving and vibrating vocal folds during phonation. This gives much more information about the individual vocal fold status. Video recording is done and studied in detail. A printed report with the images is given to the patient.

**Advantages:**

- Differentiating between structural and functional causes of phonatory gap
- Early detection of recovery of vocal fold paralysis and decision regarding further treatment
- Early detection of carcinoma in situ of glottis

Our Stroboscope facility is the first of its kind in central Kerala, and fourth in the State.

**B] Voice analysis and therapy**

In the Audiology Dept., the patient’s voice and speech are recorded, documented and analyzed. Subjective and objective assessments to check different parameters such as F0, Jitter, Shimmer, Glottic closure quotient etc. are done. These are also given as printouts to the patient.

Therapy procedures include pitch training (for Puberphonia), loudness training (for Phonatory gap), articulatory feedback treatment (for Cleft and other oral cavity defects) and phonation duration improvement. The Dr. Speech software is of immense use in providing behavioural feedback for the patient, which is an important part of voice therapy.

**Audit**

After the initial hiccups and learning curve, we are now managing all the voice cases satisfactorily. As a part of this progress, we decided to conduct an audit of this Clinic. Following is a brief round-up of the patient load in our Voice Clinic, during the period January 2012 to December 2012.

- Total patients seen: 158
- New cases: 59
- Stroboscopy: 46
- Voice therapy:
  - More than 350 sessions for the above patients

**Phonosurgeries:** Microlaryngoscopy for benign vocal fold lesions: 48 Thyroplasty Type 1 (medialization of paralysed vocal fold): 1
- Fat injection (for sulcus vocalis): 1
- Glottic web excision: 3

**Results:** A detailed audit of post-treatment results would be out of scope of the present review. Approximately 90% of our patients have reported satisfactory resolution of their voice issues. Subjectively, VHI scores, and objectively, stroboscopy findings and Dr. Speech parameters have improved demonstrably in these cases.

**Future possibilities and Vision:**

Increasing awareness about the Voice Clinic
Upgradation of equipment, especially research-oriented
Performing more phonosurgery procedures
Conferences and workshops related to Voice and Phonosurgery
Premier centre for Voice
The impact of a dedicated multidisciplinary, comprehensive care team on patient perceptions and quality of life in chronic pancreatitis


ABSTRACT

Background: Chronic pancreatitis is a lifelong illness with remissions and relapses. Attacks of pain, steatorrhea and resultant malnutrition, diabetes mellitus with its complications and the looming risk of pancreatic cancer make life miserable for the victims. Frequent medications, hospitalizations and surgeries for pain or local complications may be required. Added to this is the psychological and financial impact on the family. The prospect of decreased longevity and low quality of life drive these patients to despair.

Chronic pancreatitis is a classic example of a chronic incurable illness in which a positive intervention by a multidisciplinary team of committed health providers can be very effective. The working of such a team in a tertiary referral centre in India is described.

Material and Methods: One hundred consecutive adult patients who have been attending the pancreas clinic of our hospital for 2-6 years voluntarily answered a structured questionnaire. Self-reported symptomatic improvement, reduction in use of analgesics, perception of disease, diabetic control, overall well-being, reduction in use of alcohol and smoking and improvement in health-related quality of life were analyzed.

Results: There was significant improvement in symptoms, subjective perceptions, overall health, diabetic state, quality of life and reduction in use of analgesics and use of alcohol and smoking.

Conclusion: A proactive multidisciplinary team can make a definite impact on the overall well-being of chronic pancreatitis patients.

INTRODUCTION

Frequent hospitalizations, loss of earning power and the financial strain imposed on the patient and family alike are other intangible factors that impact on the long-term outcome of the disease. Additionally, in alcoholic subjects the familial, social and financial disruptions adversely impact the quality of life.

Modern hospital-based care of chronic illness tend to revolve around efforts to offer palliation and relief of physical ailments, are often medicine-centered with little time spent on providing positive counseling and emotional support. This gap has to be bridged. There is a current view that individual consultations should be substituted with multidisciplinary professional teamwork, with optimal collaboration and coordination between professionals in the delivery of integrated care for the provision of high quality care. This requires a proactive team approach with a patient cohort capable of and motivated for self-management and a culture of quality improvement in such a program of care. The goals of this team approach are providing quality health care, with the WHO definition of health in view (physical, mental and social well-being, and spiritual well-being added as an afterthought). Our pancreas clinic team decided to discard the conventional passive or negative perception of an incurable chronic disease for a more positive one of confidence and hope.

AIM OF THE STUDY

To assess the impact of a multidisciplinary comprehensive pancreatitis care team on improving symptoms, patient well-being, perceptions of the disease and quality of life in patients with chronic pancreatitis.

The effect of this approach on patients’ abstinence from alcohol and tobacco was also assessed.

MATERIAL AND METHODS:

One hundred consecutive patients of CP, aged >18 years, who had been prospectively followed up for 2 to 6 years in the pancreas clinic of the Amrita Institute of Medical Sciences, Kochi, India, a tertiary referral hospital, were included in the study. Diagnosis of CP was confirmed by calcification or typical changes on imaging (ultrasonography, computerized tomography, endoscopic retrograde pancreatography, magnetic resonance imaging or endosonography). There were 66 males and 34 females (Table 1). Baseline characteristics of patients are shown in Tables 2 and 3. Thirty-four

| Table 1: Gender of chronic pancreatitis patients attending pancreas clinic (n = 100) |
|-----------------|--------------|-------|
| Gender          | Number       | Percent |
| Male            | 66           | 66.0   |
| Female          | 34           | 34.0   |
Amrita Journal of Medicine

were alcoholic chronic pancreatitis and 66 idiopathic chronic pancreatitis patients. Patients who had major complications of CP necessitating surgery or with complicating pancreatic cancer were excluded.

Each subject was required to complete a questionnaire at the time of entry and at the end of 2-6 years of attending the clinic, designed to assess changes in their perceptions of the disease, symptom improvement, quality of life, abstinence from alcohol and smoking, over-all well being and self-assessment of the degree of benefit he/she derived, if any, from attending the pancreas clinic.

The health-related quality of life was assessed based on a modified and validated SF-36 questionnaire.6

Written informed consent was obtained from each subject. Approval of institutional ethics committee was obtained prior to the study which conformed to international ethical guidelines.7

Statistical methods used in analysis of data

Statistical analysis was done using SPSS (Version 11) Software Package.

Design and functioning of the comprehensive care pancreas clinic

In what way did our approach to patient care differ from the conventional?

The core principle of our project is to make our care “patient-centered” rather than “medicine-centered”. We shifted the emphasis on regular frank discussions on the nature of the illness, and the rationale for all investigations and treatment we were advising, with patient as well as immediate care-giver. Secondly, during each follow-up visit we were particular to offer positive counseling on emotional issues along with counseling on diet and life-style. Subjects are encouraged to discuss with us their familial and job-related problems to which we try to offer helpful suggestions. This helps the patient to develop confidence and rapport with each team member. Each visit is utilized to drive home the importance of abstinence from alcohol and tobacco. Medicines and procedural interventions are optimally utilized, after explaining their benefits and possible adverse effects. Every decision about investigations, drugs and procedures is taken by the team members considering patient preferences and economic viability. Surgical procedures are undertaken only when the physician, along with surgeon and patient, concurred on such decision. A central point of our approach was that the patient was accepted as an important partner in his treatment.

The core team consisted of two gastroenterologist physicians with focused interest in pancreatic diseases,

![Figure 1: Algorithm showing path of work up, management and follow up of patients attending the Pancreas Clinic at Amrita Institute of Medical Sciences and Research Center, Cochin, India.](image)

<table>
<thead>
<tr>
<th>Educational level</th>
<th>Number</th>
<th>Percent</th>
</tr>
</thead>
<tbody>
<tr>
<td>School</td>
<td>53</td>
<td>53.0</td>
</tr>
<tr>
<td>Graduate</td>
<td>38</td>
<td>38.0</td>
</tr>
<tr>
<td>Postgraduate</td>
<td>6</td>
<td>6.0</td>
</tr>
<tr>
<td>Professionals</td>
<td>3</td>
<td>3.0</td>
</tr>
</tbody>
</table>

| Characteristics of patients on enrollment in the pancreas clinic (n = 100) |
|------------------|--------|---------|
| Number            | Percentage |
| Pain              | 97     | 97.0    |
| Diabetes          | 64     | 64.0    |
| Alcohol abusers   | 34     | 34.0    |
| Regular smokers   | 30     | 30.0    |

Table II: Educational level of chronic pancreatitis patients (n = 100) (n = 100)
The First Visit

A full work up of the patient is completed, with history and physical examination, by the physician during the first visit, after registration. Further, the physician discusses about the illness with the patient and primary care giver, and answers their queries and addresses their anxieties. He briefly advises them regarding diet and lifestyle changes and then prescribes drugs to be taken regularly and those to be taken during episodes of pain. All data are stored and maintained in an electronic database. Further, investigations are ordered, referrals arranged and appointments scheduled. Quality of life assessment is done in all patients to identify patients with impaired physical and mental scores, and to plan an intensive program of medical and psychological follow-up. The path followed in work-up and follow-up of patients is given in the algorithm (Figure 1).

Dietary assessment is done by a trained dietician using food frequency and 24-hour diet recall, followed by dietary counseling and chalking out a dietary plan on an individual basis. The dietary plan is aimed at maintaining an optimal weight, as some patients are underweight and some others obese.

Current alcohol intake, smoking and use of other forms of tobacco, including frequency, duration and quantity of use, and history of life-time usage, along with other risk factors such as drug intake are elicited from the patient by personal interview by a trained medical social worker, using lifestyle assessment questionnaire, further verifying the data from the care-giver.

Patients are provided a free booklet on pancreatitis containing information on the disease, its likely risk factors, manifestations, treatment, course and prognosis in a simple question-answer format in the local tongue (Malayalam) or in English.

A fast-track card with names and telephone numbers and e-mail IDs of contact persons in the group is provided; any one of the team members can be contacted for advice in case of an emergency.

Follow-up visits

Follow-up visits are scheduled once in 3-6 months during which data on current weight, presence and grade of severity of pain, steatorrhea, diabetic status, drug intake, complications, and global well-being are recorded on a structured pro-forma. Details of dietary habits and patterns, and lifestyle are verified and recorded. Hazards of continued alcoholism and smoking are re-emphasized during these visits to patient and caregiver by medical sociologist, further endorsed by the physician. Early evidence of malignancy is monitored by suspicious clinical symptoms and signs, tumour markers, ultrasound/CT every 6 months to 1 year.

A patient support group, SHARE (Sharing Health Awareness, Research and Education) is functioning and meets periodically with the team support.

Pain has a deep emotional, subjective component, which, when attended to, raises pain threshold and helps to decrease its intensity and frequency. Patients use less of analgesics and narcotics when they regularly attend the clinic, and it is our impression that even the need for surgical intervention for pain relief has come down.

Even though a recent Cochrane Review observes that complex interventions aimed at enhancing patient adherence to medication do not work as effectively in long-term health care as in acute illnesses, 10 perhaps due to our proactive team approach, we have observed good compliance with medication and instructions.

Patients are encouraged to lead an active life with continuation of all their normal activities and family life. Physical mobility and activity are encouraged. Regarding the levels of physical exercise that are permitted, our advice will be that each one finds his/her own limits. We educate the patients on the basis for their symptoms and encourage them to identify precipitating factors and to find preventive solutions themselves.

**RESULTS**

Assessment of the impact of the pancreas clinic on patients’ symptoms, lifestyle, quality of life and subjective perceptions.

Many of the outcomes that accrue for patients attending the pancreas clinic are not measurable. However, we attempted to assess in a graded manner, the benefits

<table>
<thead>
<tr>
<th>Parameters</th>
<th>Very much reduced</th>
<th>Somewhat reduced</th>
<th>No change</th>
<th>Worse/More</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pain (n=97)</td>
<td>86 (88.6%)</td>
<td>-</td>
<td>11 (11.3%)</td>
<td>-</td>
</tr>
<tr>
<td>Use of analgesics (n=97)</td>
<td>60 (61.8%)</td>
<td>27 (27.8%)</td>
<td>4 (0.04%)</td>
<td>6 (0.06%)</td>
</tr>
<tr>
<td>Use of Narcotics (n=19)</td>
<td>6 (31.5%)</td>
<td>11 (57.8%)</td>
<td>0</td>
<td>2 (0.1%)</td>
</tr>
<tr>
<td>Diabetes (n=64)</td>
<td>26 (40.6%)</td>
<td>28 (43.75%)</td>
<td>6 (0.09%)</td>
<td>4 (0.06%)</td>
</tr>
<tr>
<td>Feeling of Indigestion</td>
<td>61 (61%)</td>
<td>31 (31%)</td>
<td>5 (5%)</td>
<td>3 (3%)</td>
</tr>
</tbody>
</table>
that our patients derive by attending the pancreas clinic regularly. The parameters we assessed were improvement in pain and other symptoms, reduction in use of analgesics, control of diabetes, changes in subjective perceptions of the disease, abstinence from alcohol and smoking, and health related quality of life (HRQoL). The demographic characteristics of the patients and their responses to the questionnaire are given below (Tables I to VI). A scoring system was devised to grade the degree of improvement. Weighted scores were assigned to 5 of the most important parameters using a scale ranging from −1 to +2 for four of these and scale of 0 to +2 for the fifth. Using this scoring, the maximum possible score of improvement a patient could achieve was +10 while maximum score for worsening was −4. It was seen that 67 % of subjects had good score, 23% had average score, while only 8% and 2% had poor score and worsening (negative score) respectively (Figure 2).

In addition, (HRQoL) was also calculated, using Short Form-36 (SF-36) on enrollment and regular follow-up in the clinic for 2 to 6 years (Figure 3).

**DISCUSSION**

The principles underlying a comprehensive approach to a chronic debilitating illness are by no means new. A Cochrane Collaboration review showed that comprehensive interventions in the form of combination of health care provider education, organizational changes in the personnel or management of visits and follow-up, computerized tracking and reminder systems, and organized approaches to follow-up helped improve the primary care of diabetic patients. Various other workers have also attempted to develop a Chronic Care Model (CCM) to improve the care of patients with chronic illness. An important consideration is to attempt to address human needs in a graded manner as espoused in Maslow’s hierarchy of needs in order to achieve optimum levels of motivation and action.

Our goal was to provide integrated care so as to meet the WHO definition of Health, which includes physical, mental and social well-being, with spiritual well-being added later.

Our evaluations revealed that the pancreas clinic bestowed substantial improvement in symptoms, lifestyle and subjective perceptions of patients with CP after 2 to 6 years of follow-up in the pancreas clinic. There was significant improvement in HRQoL assessed using the SF-36. These improvements were attributable to several factors: reduction in the subjective component of pain, better compliance, psychological support, quicker intervention during worsening of pain or onset of complications, modification of diet and lifestyle, and finally, through patient and bystander empowerment. A recent report suggests that patient-physician connectedness may affect quality of primary care.

Since the aims of dietary advice given to patients were directed to maintain an ideal weight, rather than weight gain, it was found that many underweight and overweight patients could achieve this target.

The high rates of abstinence from alcohol in our patient group must have contributed significantly to symptom improvement in ACP patients. This was achieved by regular and repeated counseling and motivation. The impact of repeated interventions in increasing abstinence rates from alcoholism has been recently highlighted in an editorial in Gastroenterology. In the same issue of the journal, a study reports reduction in recurrence rates of recurrent acute pancreatitis by

**Table V. Alcohol abuse and smoking on enrollment in the pancreas clinic and at > 2 years follow up**

<table>
<thead>
<tr>
<th>Disease Risk factors</th>
<th>On enrollment (n &amp; %)</th>
<th>Abstaining at 2 years or more of follow up (n &amp; %)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Alcohol abusers (Adult males)</td>
<td>34/62 males (54%)</td>
<td>31/34 (91%)</td>
</tr>
<tr>
<td>Regular Smokers (Adult males)</td>
<td>30/62 males (48%)</td>
<td>23/30 (76%)</td>
</tr>
</tbody>
</table>
repeated (six monthly) counseling for alcoholism.\textsuperscript{15} It is for the first time, that a significant increase in abstinence rates along with improvement in symptoms and overall health of patients are being reported as a result of repeated (every 3 to 6 months) motivational counseling in CP patients (during a 2-6 year period).

In addition, we have been able to achieve a substantial cessation of smoking among chronic pancreatitis patients, through regular 3-6 monthly counseling. The role of smoking as an independent high-risk factor for pancreatic inflammation has recently been demonstrated in a very large Dutch cohort.\textsuperscript{16} Another recent report highlights that intensive counseling against smoking, when combined with pharmacotherapy, achieved significantly better results than pharmacotherapy alone in smoking cessation.\textsuperscript{17} Cessation or reduction in smoking might well contribute to pain reduction. However a follow up period of only 2-6 years and a cohort of only one hundred patients are insufficient to draw conclusions regarding complicating pancreatic cancer. Our experience is in agreement with Glasgow et al that long-term preventive interventions can be successfully incorporated into chronic care models.\textsuperscript{18}

Patients are trained and supported to live their lives to their best potential and lead active and productive lives, despite the illness. The importance of the right mental attitude when facing an incurable illness is elegantly expressed by Randy Pausch, facing imminent death from pancreatic cancer, when he reflects on his Last Lecture. “Many people might expect the talk to be about dying. But it had to be about living”.\textsuperscript{19} Thus the concept we present may be a model for many chronic illnesses where a conventional cure may not be achievable, but a near normal life may be within the reach of patients.

Conclusions

A pancreas clinic with a committed team offering “patient-centered” care has succeeded not only in offering substantial relief of physical ailments, but also in providing patient and bystander education, relief of anxiety, easy access to health care givers and in promoting abstinence from alcohol and smoking. Patients are supported in developing a positive attitude, in improving their overall quality of life and in leading an active and useful life. This could be a useful model of care in chronic illnesses in general.

Acknowledgements

We acknowledge the help received from Prof. K. R. Sundaram. Professor of Biostatistics, and that of T. Ranjith, V. Vidya, Asha S Nair, V. Praveena in organizing and conducting the clinic, and Dr. BN Girish, Dept of Physiology for co-ordination of research.

Conflict of Interest:

None.

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Kidney transplantation - Experience at Amrita Hospital

L. Kumar****, Sudhindran S***, Thomas A*, V. N Unni*

ABSTRACT

Background: The preferred mode of renal replacement therapy (RRT) in patients with end stage renal disease (ESRD) is renal transplantation. Kidney transplantation offers longer and better quality of life compared to dialysis. Approximately 100,000 people develop ESRD every year in India and become dialysis dependent. Renal transplant program at Amrita Institute of Medical Sciences was started in 2001.

Aim: To analyse the profile of renal transplant recipients at AIMS and to calculate the patient and graft survival.

Materials and methods: A retrospective analysis of data of all patients who underwent transplant at AIMS was done. Clinical and demographic data of donors and recipients were collected and analysed.

Results: A total of 370 renal transplants have been performed from September 2001 to March 2013. Patients from all over the state of Kerala, from various social backgrounds underwent renal transplantation at AIMS. Chronic glomerulonephritis was the leading cause of ESRD. Majority of the recipients were males and majority of the donors were females. First degree relatives constituted 81% of the donors, 14.9% were spouses and 4.1% were deceased donors. The patient survival by Kaplan Meier analysis was 96% at one year, 88% at 3 years and 78% at 10 years. Graft survival was 97% at one year, 89% at 5 years and 78% at 10 years.

Conclusion: The kidney transplantation programme at AIMS has a success rate that is comparable to international standards. With strict monitoring at every stage of the process and a proper team work, high quality kidney transplant program can be established in a low income country like India.

INTRODUCTION

The preferred mode of renal replacement therapy (RRT) in patients with end stage renal disease (ESRD), who are fit to undergo surgery, is renal transplantation. Kidney transplantation offers longer and better quality of life compared to dialysis. However, the procedure of renal transplantation is complex, as the success of transplantation is influenced by donor and recipient selection, the surgical procedure and appropriate medical management of the transplant recipient.

Renal transplant program at Amrita Institute of Medical Sciences was started in 2001. The aim of this study is to analyse the profile of renal transplant recipients at AIMS and to calculate the patient and graft survival.

MATERIALS AND METHODS

A retrospective analysis of data of all patients who underwent transplant at AIMS was done. A total of 370 renal transplants have been performed from September 2001 to March 2013. Clinical and demographic data of donors and recipients were collected and analysed. The results were tabulated. Patient and graft survival were calculated using Kaplan Meier analysis.

RESULTS

Recipients:

Out of 370 recipients, 77% were males and 23% were females (figure1). Mean age was 33 years (range: 11 to 61 yrs); median age was 30 years. The age distribution of the recipients was as follows: 11 to 20 years: 11%, 21 to 30 years: 39%, 31 to 40%: 27%, 41 to 50 years: 15%, 51 to 60 years: 7.7% and 61 to 70 years: 0.3% (figure 2). We have had patients from all parts of Kerala, and the geographical distribution is given in Table 1.

The renal transplant recipients at AIMS were from a varied social background (Table 2). One hundred and fifty three (41.4%) of the patients had O blood group, 108 (29.2%) had A group, 86 (23.2%) had B group and 23 had AB group (figure 3).

Figure 4 shows the distribution of native kidney disease. Majority (27.6%) of the recipients had chronic glomerulonephritis, 11.6% had IgA Nephropathy, 8.1% had diabetic nephropathy and 7.8% had chronic interstitial nephritis. In 23.3% of the patients, inspite of diagnostic work up, a primary cause for the renal failure could not be made out; renal biopsy was not done in these patients as kidneys were small and showed significant chronicity on ultrasonography. Hence these patients were classified as ‘unknown’ native kidney disease.

Donors:

In the donor group, 71.4% were females and 28.6% were males (figure 1). The median age of the donors was 45 years, and range was 18 to 69 years and the median age was 46 years. The age distribution of the donors was as follows: 11 to 20 years: 1.9%, 21 to 30 years:
Table 1: Geographical distribution of recipients - (District – wise)

<table>
<thead>
<tr>
<th>District</th>
<th>Number of Patients</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>Thiruvananthapuram</td>
<td>20</td>
<td>5.4%</td>
</tr>
<tr>
<td>Kollam</td>
<td>43</td>
<td>11.7%</td>
</tr>
<tr>
<td>Pathanamthitta</td>
<td>31</td>
<td>8.4%</td>
</tr>
<tr>
<td>Idukki</td>
<td>32</td>
<td>8.6%</td>
</tr>
<tr>
<td>Kottayam</td>
<td>13</td>
<td>3.5%</td>
</tr>
<tr>
<td>Alappuzha</td>
<td>42</td>
<td>11.3%</td>
</tr>
<tr>
<td>Ernakulam</td>
<td>46</td>
<td>12.4%</td>
</tr>
<tr>
<td>Thrissur</td>
<td>39</td>
<td>10.7%</td>
</tr>
<tr>
<td>Palakkad</td>
<td>13</td>
<td>3.5%</td>
</tr>
<tr>
<td>Malappuram</td>
<td>14</td>
<td>3.7%</td>
</tr>
<tr>
<td>Kozhikkode</td>
<td>23</td>
<td>6.2%</td>
</tr>
<tr>
<td>Wynad</td>
<td>03</td>
<td>0.8%</td>
</tr>
<tr>
<td>Kannur</td>
<td>32</td>
<td>8.6%</td>
</tr>
<tr>
<td>Kasargode</td>
<td>14</td>
<td>3.8%</td>
</tr>
<tr>
<td>Outside Kerala</td>
<td>5</td>
<td>1.3%</td>
</tr>
</tbody>
</table>

Table 2: Occupation of the recipients

<table>
<thead>
<tr>
<th>Occupation</th>
<th>Number of patients</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>Office job</td>
<td>81</td>
<td>22.1%</td>
</tr>
<tr>
<td>Self employed</td>
<td>75</td>
<td>20.4%</td>
</tr>
<tr>
<td>Student</td>
<td>59</td>
<td>16.1%</td>
</tr>
<tr>
<td>Small job</td>
<td>44</td>
<td>11.9%</td>
</tr>
<tr>
<td>Homemaker</td>
<td>41</td>
<td>11.0%</td>
</tr>
<tr>
<td>Manual labour</td>
<td>21</td>
<td>5.7%</td>
</tr>
<tr>
<td>Unemployed</td>
<td>15</td>
<td>4.0%</td>
</tr>
<tr>
<td>Teacher</td>
<td>14</td>
<td>3.7%</td>
</tr>
<tr>
<td>Business</td>
<td>13</td>
<td>3.4%</td>
</tr>
<tr>
<td>Agriculturist</td>
<td>7</td>
<td>1.7%</td>
</tr>
</tbody>
</table>

6.5%, 31 to 40%: 18.6%, 41 to 50 years: 40.8%, 51 to 60 years: 28.6% and 61 to 70 years: 3.2%. (Figure 2). Of the donors, 50.5% were parents, (11.6% were fathers and 38.9% were mothers); 30.5% were siblings, in whom brothers and sisters were 14.9% and 15.7% respectively; 14.9% were spousal donors and 4.1% were cadaveric donors (Figure 5).

Cyclosporine, Azathioprine and Prednisolone was the immunosuppressive regime in 41.9% of our patients. Cyclosporine, Mycophenolate mofetil and Prednisolone was given for 29%, Tacrolimus, Mycophenolate mofetil and Prednisolone was given for 24%; Tacrolimus, Azathioprine and Prednisolone was given for 2.2% of the transplant recipients.

The patient survival by Kaplan Meier analysis was 96% at one year, 88% at 5 years and 81% at 10 years (figure 6, Table 3). Graft survival was 97% at one year, 89% at 5 years and 78% at 10 years (figure 7, Table 4).
Cause of death among renal transplant recipients were as follows: Graft failure and end stage renal disease in 13 patients, pneumonia in 13, and sepsis in 13. Five patients died of other causes such as coronary artery disease and road traffic accident. Ten year graft survival was 78% in our study (Table 4).

**DISCUSSION**

The first successful kidney transplant was performed in Boston on 23 Dec 1954 between identical twins. In the early years, kidney transplantation was considered to be a risky experimental procedure with high failure rates and was limited to certain academic institutions in the developed nations. Over the decades, with the advancement of surgical techniques and the use of novel immunosuppressants, kidney transplantation has become a routine procedure even in developing countries. Over 100,000 new patients develop ESRD in India every year. The first kidney successful kidney transplantation in India was done at Vellore in 1971. Currently around 5000 kidney transplant are done per year in India.

The renal transplant programme at Amrita Institute of Medical Sciences was started in 2001 and since then 370 patients have undergone kidney transplant surgery till March 2013. Majority of the recipients were males while most of the donors were females. The donors were all first degree relatives or spouses. Of these, 355 were live related donor kidney transplantation and 15 were deceased donor transplants. Unlike the western world, where most of the transplants are deceased donor transplants, it is the live donor transplant programme which is more common in India. Among the live related donors, more than half were parents and mothers constituted 39% of all donors. This is a reflection of the sociocultural background of this region. Siblings constituted 30.5% of the total transplant donors and spouses constituted 14.9% of donors.

Chronic glomerulonephritis was the most common cause of ESRD in our study group. The primary kidney disease was unknown in 23.3% of the individuals. IgA nephropathy and diabetic nephropathy were the other major causes leading to ESRD. With the prevalence of type 2 diabetes reaching epidemic proportions in India, diabetic nephropathy has become the most common cause of ESRD in India among those aged more than 40 years. In the current study 8.6% of the recipients had diabetes mellitus.

The patient survival at AIMS was 96% at one year, 88% at 5 years and 81% at 10 years. These results are on par or even better than the international survival rates (Table 3). The Organ procurement and transplantation network (OPTN) - Scientific registry of transplant recipients (SRTR); the United States department of health and human services data showed that the 10 year survival among live donor transplant recipients was 78.8%. Ten year graft survival was 78% in our study compared to 62% in the OPTN- SRTR.

Current evidence suggests that kidney transplantation is associated with reduced risk of mortality and cardiovascular events as well as better quality of life.

### Table 3: Patient survival at AIMS. Kochi in comparison with Organ procurement and transplantation network (OPTN) - Scientific registry of transplant recipients (SRTR) - US Department of Health and Human Services data (2011)

<table>
<thead>
<tr>
<th>Duration</th>
<th>AIMS, Kochi</th>
<th>OPTN/ SRTR</th>
</tr>
</thead>
<tbody>
<tr>
<td>1 year</td>
<td>96%</td>
<td>99%</td>
</tr>
<tr>
<td>5 year</td>
<td>88%</td>
<td>93%</td>
</tr>
<tr>
<td>10 year</td>
<td>81%</td>
<td>79%</td>
</tr>
</tbody>
</table>

### Table 4: Graft survival at AIMS, Kochi in comparison with Organ Procurement and Transplantation Network (OPTN) - Scientific Registry of Transplant Recipients (SRTR) - US Department of Health and Human Services data (2011)

<table>
<thead>
<tr>
<th>Duration</th>
<th>AIMS, Kochi</th>
<th>OPTN/ SRTR</th>
</tr>
</thead>
<tbody>
<tr>
<td>1 year</td>
<td>97%</td>
<td>97%</td>
</tr>
<tr>
<td>5 year</td>
<td>89%</td>
<td>84%</td>
</tr>
<tr>
<td>10 year</td>
<td>78%</td>
<td>62%</td>
</tr>
</tbody>
</table>
when compared to maintenance dialysis. These findings were true for different dialysis modalities and for transplantation from both deceased and living donors and across countries with different health care systems. An earlier study from our Institute had shown that renal transplant recipients had a significantly better quality of life compared to those on hemodialysis. Six transplant recipients from our hospital had successful pregnancies. Most of our transplant recipients were working in either government or private sector and majority of them were able to resume their work after transplantation.

The United States department of heath and human services data (OPTN- SRTR- Annual report 2011) provides the total number of kidney transplants in different countries. The total number of kidney transplants in India was 5000 in the year 2010, which included 4900 live donor and 100 deceased donor transplants and this number translates to transplant rate of 4.26 per one million. This is extremely low compared to countries like Norway and the United States where the rates are 59.2 and 57.5 per million respectively. Although this mirrors the socio-economic and cultural differences between India and the wealthy western nations, there is a definite scope for improvement in our country.

In conclusion, at AIMS, Kochi we have patients from all over the state of Kerala, from various social backgrounds undergoing renal transplantation. The kidney transplantation programme at AIMS has survival rates comparable to international standards. Our transplant recipients also have a significantly better quality of life than those on dialysis. With strict monitoring at every stage of the process and a proper team work, high quality kidney transplant programme can be established in a low income country like India. The need of the hour is mass awareness programmes on organ donation, more centres with infrastructure and expertise for organ transplantation and overall, a positive move from the general public as well the governmental bodies for promoting organ donation.

Acknowledgement

The authors would like to thank the faculty of department of Pathology, Microbiology, Biochemistry, Radiology, Nuclear medicine, Cardiology, Gastroenterology, Gynecology, ENT, Dental surgery, Ophthalmology and Psychiatry as well as the nursing and technical staff in dialysis unit and post transplant ICU for their help and cooperation in the care of our kidney transplant recipients.

We would also acknowledge the sincere and dedicated efforts of the residents, the transplant coordinator, the medical social worker, the physician assistants and the administrative staff of department of Nephrology.

REFERENCES

Ifosfamide is a synthetic analog of Cyclophosphamide that has been approved for concurrent use with other drugs (usually Cisplatin, Etoposide or Vinblastine) in the treatment of sarcomas and metastatic germ-cell testicular cancer. Nephrotoxicity due to direct tubular injury is a prominent complication of Ifosfamide therapy. In addition to tubular dysfunction, reduction in glomerular filtration rate is generally mild unless ifosfamide is given in combination with another nephrotoxin such as Cisplatin.

CASE REPORT

A 21 year male presented in December 2009 with hemoptysis, dull aching diffuse chest pain and progressive dyspnea of one month duration and bilateral multiple rounded lung parenchymal lesions on X-ray chest (Figure 1a); he was diagnosed to have primary synovial sarcoma on histology (CT Guided Lung biopsy) and immunohistochemistry was positive for vimentin, B cell lymphoma - 2(Bcl-2), calretinin and CD 99. He was initially treated with chemotherapy [Adriamycin - 160mg plus Cisplatin 120mg (6 cycles) followed by Gemcitabine plus Docetaxel (3 cycles)] over a period of 9 months till September 2010. He had symptomatic relief and resolution of pulmonary lesions on X-ray.

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creatinine 4.7 mg/dl, eGFR 16.8 ml/min/1.73 m²). He was initiated on hemodialysis and renal biopsy was performed. Renal histology showed predominantly tubulointerstitial changes (simplification of the epithelium, interstitial edema and moderate lymphocytic and plasma cell infiltrates and interstitial fibrosis with tubular atrophy - 20%). Few tubules showed atypically enlarged tubular cells with great variation in size and shape of the nuclei, mostly with visible nucleoli (figure 2b). Glomeruli appear near normal in size and cellularity. Peripheral capillary loops, interlobular arteries and arterioles appear unremarkable. Hemodialysis was continued and he is dialysis dependant for the last 12 months.

**DISCUSSION**

Synovial sarcomas accounts for approximately 8% of soft tissue sarcomas (2). These tumors are not derived from the synovium, but from immature mesenchymal elements. They are divided into four histologic types: biphasic, monophasic fibrous, monophasic epithelial, and poorly differentiated. Our case was a poorly differentiated type. Diagnosis is made from histology, supplemented with immunohistochemistry (IHC) and cytogenetic studies. Immunohistochemically, synovial sarcomas are nearly uniformly positive for cytokeratin, EMA (Epithelial Membrane Antigen), Bcl-2, and vimentin, and negative for (Solubility in 100% dilution)S-100, desmin, smooth muscle actin, and vascular tumor markers3. IHC studies in this case were positive for vimentin, Bcl-2, calretinin and CD 99. Cytogenetic studies of synovial sarcomas have revealed the chromosomal translocation t(x;18) (p11;q11). This translocation fuses the SYT gene from chromosome 18 to either of two homologous genes at Xp11, SSX1 or SSX2. SYT-SSX1 and SYT-SSX2 are thought to function as aberrant transcription regulators. The sensitivity of this test for diagnostic purposes approaches 100%2.

Synovial sarcoma typically presents in adolescents and young adults (average age at presentation 25 yrs), most commonly in the soft tissues of the extremities, especially near large joints, but other sites like head and neck, lung, heart, mediastinum, and abdominal wall have been reported. Synovial sarcoma arising from the lungs and pleura have rarely been reported. Pulmonary sarcomas are very uncommon and comprise only 0.5% of all primary lung malignancies3.

The prognosis for patients with pulmonary synovial sarcoma is poor, with an overall 5-year survival rate of 50%. Factors predicting a worse prognosis include tumor size (> 5 cm), male gender, older age (> 20 years), extensive tumor necrosis, high grade of malignancy, large number of mitotic figures (> 10 per 10 high-powered fields), neurovascular invasion, and, recently, the SYT-SSX1 variant [3]. The main prognostic factor is the ability to achieve a complete resection. The prognosis for patients with the SYT-SSX2 abnormality is better (no deaths in the first 5 years after surgery in one study group) than that for patients with the SYT-SSX1 abnormality4.

There is no standardized therapy; most patients are treated with surgery or with surgery and adjuvant radiation therapy. The rarity of this tumor has not permitted controlled studies on adjuvant chemotherapy. Synovial sarcomas are chemosensitive and the agents tried include Cisplatin, Ifosfamide, Doxorubicin, Gemcitabine and Docetaxel, with an overall response rate of approximately 24%4.

Renal dysfunction related to treatment of synovial sarcoma

**Cisplatin**

Cisplatin is a common antineoplastic drug used for the treatment of solid tumors. Its chief dose limiting side effect is nephrotoxicity; 20% of patients receiving high-dose cisplatin have severe renal dysfunction5. The kidney tissue accumulates Cisplatin to a greater degree (5 times the serum concentration) than other organs and is the major route for its excretion. However, conversion of Cisplatin to nephrotoxic molecules in the proximal tubule cells is necessary for cell injury. Cisplatin is conjugated to glutathione and then metabolized through a gamma glutamyl transpeptidase and a cysteine
S-conjugate β-lyase–dependent pathway to a reactive thiol, a potent nephrotoxin. Cisplatin nephrotoxicity can present in a number of ways. Acute toxicity is mainly reflected by proximal tubular dysfunction. Acute cisplatin nephrotoxicity is dose-dependent, but can be largely prevented by adequate hydration of the patients. The renal insufficiency, typically non-oliguric, begins several days after the dose of Cisplatin, and the urine may contain glucose and small amounts of protein. Recovery of renal function usually occurs over a period of 2–4 weeks after stopping the drug. Progressive and permanent nephrotoxicity can result with successive treatment courses despite preventive measures. In contrast to the good outcome of acute nephrotoxicity, controversy exists regarding the deleterious long-term renal side-effects of this agent.

Cisplatin-induced renal injury probably does not have any specific clinical features. Urinary excretion of a proximal tubular injury markers, such as β-2 microglobulin, N-acetyl-Times New Roman-D-glucosaminidase, and Times New Roman1-α-acid glycoprotein, increase after cisplatin treatment. There is little change in urine protein excretion. Renal histology is characterized predominantly by tubular alterations, such as atypically enlarged tubular cells with great variation in size and shape of the nucleus, mostly with visible nucleoli. In the interstitium, infiltration of mononuclear leukocytes is seen. Electron microscopy shows no major glomerular abnormalities.

There is no specific treatment for cisplatin-induced renal dysfunction or injury. These patients need careful attention to hydration and management of electrolyte disturbances. Cisplatin induces magnesium depletion, and magnesium deficiency itself may enhance Cisplatin nephrotoxicity. Therefore, magnesium repletion may attenuate cisplatin-induced nephrotoxicity. Other agents being tried in prevention of Cisplatin nephrotoxicity include antioxidants, Procainamide, salicylates, and use of newer drugs like Carboplatin and Oxiplatin. In addition, these patients should avoid, to the extent possible, other nephrotoxic agents, including intravenous radiographic contrast and nephrotoxic antibiotics.

Ifosfamide

Ifosfamide is a structural analog of Cyclophosphamide. However, despite the structural similarities, Cyclophosphamide and Ifosfamide have important differences in their metabolism, toxicity, and therapeutic spectrum. Approximately 45% of the therapeutic dose of Ifosfamide is typically metabolized via N-dechloroethylation to chloroacetaldehyde (CAA), whereas only 10% of Cyclophosphamide is converted to CAA. As CAA is thought to induce neurotoxicity and nephrotoxicity, this is likely to account for the higher prevalence of these untoward events among patients treated with Ifosfamide. Specific toxicities related to Ifosfamide include hemorrhagic cystitis (caused by acrolein and can be prevented with mesna), neurotoxicity and nephrotoxicity.

The incidence of renal toxicity varies between 5% and 30% and CAA is proposed to be the predominant nephrotoxin and mesna has no effect on it. Although renal damage is often acute and reversible, chronic toxicity may develop with potentially serious consequences. Proximal tubular dysfunction is the commonest presentation, and may lead to a Fanconi syndrome, including hypophosphataemic rickets and proximal renal tubular acidosis; although distal tubular impairment has been described, it is relatively rare.

Several risk factors for the development of chronic nephrotoxicity have been described; total Ifosfamide dose, age of the patient, previous or concurrent Cisplatin treatment, and unilateral nephrectomy are the most important risk factors. Brandis estimated the overall incidence of renal tubular dysfunction in patients treated with Ifosfamide to vary between 10 and 20%, with only 1-3% of cases showing severe clinical symptoms.

Chronic renal failure due to Ifosfamide is extremely rare and occurs in patients treated with very high doses (daily dosages >5 g/m2). There are no known measures to prevent Ifosfamide renal toxicity.

Because of the ongoing dispute and the lack of consensus on the best chemotherapeutic regimen, numerous Ifosfamide regimens (both as monotherapy and in combination with others) have been tried in patients with soft tissue sarcomas. Monotherapy showed a response rate of 38% and median overall survival of 1 year. Several trials have been performed exploring the feasibility of Ifosfamide-containing combinations and whether or not such combinations are more effective than single-drug regimens. The combinations studied are Doxorubicin plus Ifosfamide (high response rates of 50%–60%), four-drug combination CYVADIC (Cyclophosphamide, Vincristine, Doxorubicin, and Dacarbazine), Ifosfamide plus Paclitaxel and Gemcitabine plus Docetaxel.

Nephrotoxicity can be documented few months after completion of Ifosfamide treatment and sometimes renal functions may gradually worsen. Outcome varies between individual patients; but the overall prognosis remains poor. Several risk factors like total Ifosfamide dose, age at treatment, previous or concurrent Cisplatin treatment and unilateral nephrectomy predicts the outcome.

CONCLUSION

Primary pulmonary synovial sarcoma is an extremely rare malignancy of the lungs and pleura which requires histology, IHC for the diagnosis and cytogenetic studies
for prognostication. The poor prognostic factors in our case were tumor size (>5 cm), male gender, poorly differentiated histology. The prior use of Cisplatin increased the risk of nephrotoxicity related to Ifosfamide and the use of mesna did not afford protection from nephrotoxicity. Chronic renal failure due to Ifosfamide is extremely rare and occurred in our patient although he received much lower dose (1.5-1.8g/m2) than that has been described in literature.

**REFERENCES**

71 year old male, chronic smoker, pan chewer and diabetic, presented to a local ENT surgeon with symptoms of reduced hearing on the right side. Hearing loss was associated with purulent discharge from bilateral ears, more profusely from the right side. He was advised topical ear drops, but symptoms persisted. Two weeks later, he developed cough following food intake with associated difficulty in swallowing food and water and hoarseness of voice with occasional history of nasal regurgitation and slurring of speech. Symptoms were associated with weight loss of around 14 kgs over the past 4 months. He presented to us 4 weeks after the onset of these symptoms.

On examination, he was frail with stooped posture, mask like facies and pill rolling tremor was present. Pallor was present and bilateral firm, mobile, non-tender jugulodigastric lymph nodes were palpable. MMSE revealed a score of 26/30 with presence of features suggestive of Parkinsonism. Right eye examination revealed partial ptosis, lateral gaze palsy and miosis. Sweating was normal on both sides of face. Bilateral hearing loss was present. Gag reflex and palatal reflexes were absent bilaterally. Taste in the posterior 1/3 rd of the tongue could not be examined. Right trapezius and sternocleidomastoid were weak. Tongue fasciculations were present. Tongue protrusion showed deviation to the right side. Rest of the systemic examination, including ENT examination was normal.

His investigations revealed neutrophilic leucocytosis with an ESR of 52 mm/hr and CRP of 132. His hepatic and renal function parameters were within normal limits. CT Brain showed retropharyngeal/nasopharyngeal hypodense lesion and erosion of right petrous apex. MRI Brain revealed ill defined bright signal seen diffusely involving the petrous bone, clivus in the center and extending into soft tissues and the base of skull region – suggestive of osteomyelitis of petrous and clivus with infection spreading into nasopharynx. Subsequently, a trans-nasal biopsy was obtained from the site which revealed fibrocollagenous stroma with inflammatory cell infiltrate composed of neutrophils, macrophages and lymphocytes and plasma cells. Multi nucleate giant cells were also seen with granulation tissue. Necrotic bony tissue fragments was noted amongst granulation tissue with giant cell reaction around – suggestive of chronic osteomyelitis. Anti-pseudomonal antibiotics were started empirically, but the patient succumbed to his illness.

**DISCUSSION**

Skull base osteomyelitis is a potentially life threatening infection that classically presents as a complication of severe external otitis, middle ear, mastoid or sinus infection and can result in multiple lower cranial nerve palsies as a result of jugular foramen involvement due to widespread involvement of the skull base. (1)

It is known to occur almost exclusively in diabetics and the elderly. The exact pathogenesis is not known but a flurry of factors are believed to be contributory, namely:

a. Microangiopathy

b. Hypoperfusion

c. Diminished host resistance – Impaired phagocytosis, poor leucocytic response, impaired intracellular digestion of bacteria

Pseudomonas Aeroginosa has been reported as the most common pathogen involved in skull base osteomyelitis – which has been at-
tributed to the less acidic nature and lower lysozyme content of ear wax that makes it increasingly susceptible to pseudomonas infection. It was first reported by Meltzer and Keleman in 1959. However, the term, ‘malignant otitis externa’ was first coined by Chandler who described it in 1968. It is normally described in patients with predisposing factors such as an immunocompromised state, diabetes mellitus, chronic mastoiditis, paranasal sinus infection or necrotizing otitis externa and causes progressive, unrelenting otalgia, unresponsive to local treatment.

Certain hypothesis suggest that external otitis progresses from the external auditory canal to the temporal bone and eventually the skull base via the fissure of Santorini and the osseo-cartilaginous junction. Infection enters most often by minor trauma and aural irrigation.

The presentation of skull base osteomyelitis as Villaret’s syndrome, as in our case, is extremely rare. In this syndrome, there is involvement of Cranial nerves IX, X, XI, XII and post ganglionic sympathetic fibres to eyes. The site of lesion is retropharynx, where these structures are in close proximity.

Cranial nerve involvement maybe due to both pseudomonas neurotoxins and inflammation occurring along the skull base as the disease progresses. Facial nerve is classically the most common first cranial nerve involved in the disease process, at the stylomastoid foramen. Cranial nerves IX, X and XI may become affected as the jugular foramen becomes involved, as may V and VI if the petrous apex is affected.

Levenson’s criteria maybe used to diagnose malignant otitis externa, which includes

a. Refractory otitis externa
b. Severe nocturnal otalgia
c. Purulent otorrhoea
d. Presence of pseudomonas and granulation tissue in the external auditory canal,
e. Diabetes or an immunocompromised state

Staging of the disease is generally by :

a. Stage 1 – Purulent otorrhoea, otalgia (out of proportion), granulation tissue on otoscopy
b. Stage 2 – Disease extends to soft tissues and skull base. Involvement of CN XI, XII
c. Stage 3 – Intracranial extension

Intracranial complications include meningitis, brain abscess and dural sinus thrombosis. Sigmoid sinus thrombosis should be considered if the disease involves the jugular foramen, likewise, cavernous sinus thrombosis should be considered if there is evidence of CN V or VII involvement.

Our patient had an atypical presentation, with minimal symptoms in the ear, except for the past history of discharge from bilateral ears. He presented with features of multiple lower cranial nerve palsies and sympathetic chain involvement, suggestive of Villaret’s syndrome and an additional right VIth nerve palsy, possibly due to the lesion in the petrous apex. The involvement of IX, X, XI and XII and cervical sympathetic fibres is due to the presence of lesion in retropharyngeal space, which has extended from the skull base.

The most common etiologies for Villaret’s syndrome are - parotid gland tumor, increased pressure by an enlarged lymph gland, nasopharyngeal tumors, pharyngeal abscesses and aneurysms of the internal carotid artery. Villaret’s syndrome and sympathetic chain involvement resulting from skull base osteomyelitis is a very rare presentation, and there have been very few cases reported worldwide.

REFERENCES


Villaret’s Syndrome – A Rare Presentation of Skull Base Osteomyelitis
Lung Cancer Presenting as Hyponatremia

Vishnu Dev U*, Sreedharan S***, Bindhu M.R**, Mathew A***, Rajesh R***, Kurian G***, V. N Unni***

ABSTRACT

SIADH is a disorder of water balance characterised by hypotonic hyponatremia and impaired urinary dilution in the absence of renal disease or any identifiable physiological stimulus known to release vasopressin. SIADH can develop as the result of many different disease processes that disrupt the normal mechanisms that regulate vasopressin secretion. We report a patient with SIADH due to an underlying small cell lung cancer.

INTRODUCTION

The syndrome of inappropriate antidiuretic hormone secretion (SIADH) is a condition characterised by hyponatremia and hypo-osmolality of plasma resulting from inappropriate secretion or action of the hormone leading to impaired water excretion, despite increased plasma volume1. We report an interesting case of chronic hyponatremia due to SIADH in a patient with small cell lung cancer.

CASE REPORT

A 61 year old male presented to our institution with generalised weakness and fatigue of eight months duration. He has had multiple hospital admissions in different hospitals during these eight months for generalised weakness, irrelevant talk, alteration in sensorium and two episodes of convulsions. He was found to have hyponatremia. However, the etiology remained elusive and was given parenteral 3% saline on numerous occasions. The patient also had weight loss of about 8 kgs in the last few months.

Physical examination revealed pallor and stable vital signs. Systemic Examination was unremarkable. Investigations revealed a normocytic normochromic anaemia (Hb-9.2gm%), normal leucocyte counts (8,220/ cu.mm) and platelets (2,75,000/ cu.mm). Blood sugars, renal function tests and liver function tests were normal. Urine examination did not reveal proteinuria or microscopic haematuria.

Serum potassium (3.5 mEq/L), calcium (8.1mg/dl) and uric acid (3.4mg/dl) were normal. Serum sodium was 124mEq/L, urine osmolality-753.8mOsm/kg, urine sodium-165.6mmol/l, plasma osmolality-261.8mOsm/kg and 24-hour urinary sodium was 168.8mmol/day. The thyroid function tests (free T4-1.3ng/ dl, TSH-0.5uIU/ml) and fasting cortisol (14.3ug/dl) were normal.

A diagnosis of chronic hyponatremia due to SIADH was made. ECG, X-Ray Chest and CT brain were normal. A high resolution CT scan of the chest with contrast revealed a soft tissue density mass involving the left para-aortic, para-tracheal and right hilar region, which was encasing the left pulmonary artery causing an extrinsic compression (Figure 1).

Bronchoscopy showed mucosal involvement and extrinsic compression of the left upper lobe bronchus. The mucosal surface was irregular, friable and bleeding on touch. A transbronchial biopsy was done, which revealed a small cell lung carcinoma (Figure 2a,2b). The patient was treated with Etoposide and Carboplatin. Subsequently, the sodium levels improved and the patient became symptomatically better.

Figure 1: CT chest showing the soft tissue density mass encasing the left pulmonary artery (arrow).

Figure 2a: Transbronchial biopsy of the mass showing a neoplasm composed of cells in sheets (infiltrating between native glands) with scanty cytoplasm, high N/C ratio, nuclear moulding and increased mitoses (40X).

Figure 2b: IHC for Synaptophysin (40X) – showing cytoplasmic positivity which confirms the neuro-endocrine nature.

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DISCUSSION

The syndrome of inappropriate secretion of antidiuretic hormone (SIADH) is a disorder of impaired water excretion caused by the inability to suppress the secretion of antidiuretic hormone (ADH). If water intake exceeds the urine output, the ensuing water retention leads to the development of hyponatremia.

Antidiuretic hormone (ADH or arginine vasopressin) secretion results in a concentrated urine and therefore a reduced urine volume. The higher the plasma ADH, the more concentrated the urine. In most patients with SIADH, ingestion of water does not adequately suppress ADH and the urine remains concentrated. This leads to water retention and increase in total body water. This increase in the total body water lowers the plasma sodium concentration by dilution. In addition, the increase in total body water transiently expands the extracellular fluid volume and thereby triggers increased urinary sodium excretion; this is an attempt to bring the extracellular fluid volume towards normal and further lowers the plasma sodium concentration.

Hyponatremia (serum Na+ < 135 mmol/L) with concomitant hypo-osmolality (serum osmolality < 280 mOsm/kg), high urine osmolality and high urine sodium is the hallmark of SIADH. However, these findings only indicate that ADH is present and acting on the distal nephron; it does not indicate if the ADH secretion is “inappropriate.” A good clinical examination is required to confirm that the hyponatremia is not the result of decreased effective circulating volume due to volume depletion or conditions such as congestive heart failure and cirrhosis, in which the secretion of ADH is “appropriate.” Hence renal, cardiac and hepatic dysfunction needs to be excluded before a diagnosis of SIADH is made. Hypothyroidism and Cortisol deficiency needs to be ruled out as well.

The etiology of SIADH is diverse, which can be categorised as follows 2:

1). Pulmonary Disorders
   a) Infections - Pneumonias, Tuberculosis, Pulmonary abscess, Aspergillosis
   b) Airway Diseases - Bronchial Asthma

2). Malignancies
   A). Carcinoma
      a) Lung - Small cell cancer, Mesothelioma
      b) GIT - Stomach, Duodenum, Pancreas
      c) Genitourinary tract - Ureter, Bladder, Prostate
   B). Lymphomas
   C). Sarcomas

D). Ewing’s sarcoma

3). Central Nervous System
   a) Infections - Viral Encephalitis, Meningitis, Brain abscess, Tuberculosis
   b) Intracranial bleed - subdural hematoma, subarachnoid hemorrhage
   c) Obstructive conditions - Hydrocephalus, cavernous sinus thrombosis
   d) Demyelinating disorders - Multiple Sclerosis, Guillain-Barre Syndrome
   e) Vascular events - Cerebrovascular accidents
   f) Space occupying lesions - Brain tumours

4). Drugs
   a) Drugs that stimulate release of AVP or enhance its action: Chloropropamide, Selective Serotonin Reuptake Inhibitors, Tricyclic antidepressants, Carbamazepine, Antipsychotic drugs and Nonsteroidal anti-inflammatory drugs
   b) AVP analogues - Desmopressin, Oxytocin and Vasopressin

5). Miscellaneous
   a) Hereditary (gain-of-function mutations in the vasopressin V2 receptor)
   b) Transient - administration of general anesthesia, during pain or stress.

Small cell lung cancer (SCLC), previously known as oat cell carcinoma, is considered distinct from other lung cancers, because of their clinical and biologic characteristics. Small cell lung cancer is a neuroendocrine carcinoma that exhibits aggressive behavior, rapid growth, early spread to distant sites, exquisite sensitivity to chemotherapy and radiation and frequent association with distinct paraneoplastic syndromes. The production of various peptide hormones leads to a wide range of paraneoplastic syndromes; the most common of these are the syndrome of inappropriate secretion of antidiuretic hormone (SIADH) secretion and the syndrome of ectopic adreno-corticotropic hormone (ACTH) production. Paraneoplastic syndromes are rare disorders that are triggered by an altered immune system response to a neoplasm or ectopic production of a hormone or cytokine. About 15% of small cell lung cancers are known to be associated with SIADH. Hence a patient presenting with SIADH should be fully evaluated to determine its etiology.

In the absence of a single laboratory test to confirm the diagnosis, SIADH is best defined by the classic Bartter-Schwartz criteria, which can be summarized as follows:
a) Hyponatremia with corresponding hypo-osmolality
b) Continued renal excretion of sodium
c) Urine less than maximally dilute
d) Absence of clinical evidence of volume depletion
e) Absence of other causes of hyponatremia
f) Correction of hyponatremia by fluid restriction

The treatment of SIADH and the rapidity of correction of hyponatremia depends on 6:
1) The degree of hyponatremia
2) Whether the patient is symptomatic or not
3) Whether the hyponatremia is acute (< 48 hours) or chronic.

The urine osmolality and creatinine clearance must also be considered when choosing the type of therapy. If no history is available to determine the duration of hyponatremia and if the patient is asymptomatic, it is reasonable to presume the condition is chronic. Diagnosis and treatment of the underlying cause of SIADH is also important.

In our patient, it was a small cell lung cancer that was responsible for the SIADH and chronic hyponatremia. The patient was given chemotherapy (Etoposide and Carboplatin) and subsequently, the patient has shown good clinical improvement with no further episodes of hyponatremia.

CONCLUSIONS

Chronic hyponatremia due to SIADH can be a paraneoplastic manifestation of small cell lung cancer. Symptomatic treatment alone would not correct hyponatremia. The definite etiology needs to be determined. This case highlights the importance of evaluation of hyponatremia. An interesting aspect to be considered is the presentation of small cell lung cancer in our patient. The patient presented with non-specific symptoms due to chronic hyponatremia, which could have been easily overlooked or missed. However, once the etiology was detected and treatment was given, the patient has had correction of hyponatremia.

REFERENCES

A Case Report of Neoplasia Induced Recalcitrant Lichen Planus

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INTRODUCTION

Lichen planus is a common chronic inflammatory mucocutaneous disorder which was first described clinically by Wilson in 1969 and histologically by Dubreuilh in 1906. It typically affects the oral mucosa and additionally, in some cases the skin. Lichen planus can affect other non-mucosal surfaces such as genitals, anus and pharynx. Conjunctival and oesophageal involvement is rare.

Current data suggest that OLP is a T cell mediated autoimmune disease in which auto cytotoxic CD8+ T cells trigger apoptosis of oral epithelial cells. However, the precise cause of OLP is unknown.

Although the etiology and pathogenesis of OLP are not fully understood, oral lichen planus has been associated with multiple disease processes and agents, such as viral and bacterial infections, autoimmune diseases, medications, vaccinations and dental restorative materials. It can arise in patients with autoimmune liver disease including primary biliary cirrhosis and chronic acute hepatitis. HLA DR6 may be linked to the reported association of oral lichen planus with hepatitis. Recently, several studies have reported a relationship between Helicobacter pylori and OLP. Moravej et al in 2007 found statistically significant differences in H. Pylori infection between patients with lichen planus and a control group. Genetic predisposition seems to play a role in OLP pathogenesis as several familial cases have been reported. Exacerbations of OLP have been linked to periods of psychological stress and anxiety. Altogether this makes etiology behind lichen planus a multifactorial process comprising events that may take place at different time points. Oral lichen planus affects 1-2% of the population.

The disease ranges in severity from an asymptomatic condition to very severe discomfort that may adversely have an impact on patients quality of life. Lichen planus typically arises in females of middle age but also affects males. There are yet no ethnic groups identified as being of particular risk, the condition arising in all ethnicities.

The clinical presentation is usually bilateral distribution, typically involving buccal mucosa, dorsal and ventral surface of tongue, gingiva where it presents as desquamative gingivitis. The six clinical forms of oral lesions are reticular, popular, plaque like, atrophic, ulcerative and bullous. It is usually asymptomatic, but if there are areas of ulceration, patient experiences varying degrees of discomfort, exacerbated by eating spicy or acidic foods. It presents as white patches, ulcers and rarely blisters.

The most common type is reticular form with the characteristic feature of slender white lines (Wickham’s striae) radiating from the papules. Patients with reticular lesions are often asymptomatic, but atrophic (erythematous) or erosive (ulcerative) OLP is often associated with a burning sensation and pain. A greater malignant potential has been recognized for atrophic, erosive form of OLP and the plaques form on the back of the tongue. This clinical appearance of desquamative gingivitis is not pathognomonic of erosive OLP and may represent the gingival manifestation of many other diseases such as cicatrical pemphigoid, pemphigus vulgaris, epidermolysis bullosa acquisita, and linear IgA disease.

Certain HLA types such as A3,B16,B8,BW 35 and DR1 have been detected with greater frequency than expected. Indirect immunofluorescence has at times revealed antibodies to lichen planus specific antigen. Whether the autoimmune phenomenon seen in OLP can be triggered by malignancy is genetically susceptible patients is unclear. Because paraneoplastic pemphigus can closely mimic LP both clinically and histopathologically we studied the clinical course and findings in a patient who had lichen planus following B cell lymphoma.

CASE REPORT

A 47 year old patient reported to OPD with c/o severe pain and burning sensation since 3 months. He was unable to take any food orally. He had severe discomfort and totally depressed. He was a known case of B cell lymphoma and underwent chemotherapy and radiation 3 months back following which these symptoms developed in oral cavity. No known history of drug allergy. In personal history had h/o smoking 5 cigarettes /day since 20 years and was a occasional alcoholic. On general examination he had erosive and erythematous areas in glans penis.

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On examination, patient had discoloured lips and In buccal mucosa had severe desquamation with ulcerations seen bilaterally. In lateral borders of tongue and on ventral surface also had desquamation as shown in the picture. In dorsal surface of tongue small white crops of vegetations were seen. On palpation it was severely tender and in some areas mucosa was peeling off leaving a raw eroded surface. The differential diagnosis given was pemphigus, erosive lichen planus, lichenoid reaction, mucous membrane pemphigoid and pseudomembranous candidiasis.

Investigations carried out were incisional biopsy, immunoflouresence, smear from tongue and complete blood counts. Biopsy report and immunofluoresence was confirmative of erosive lichen planus.

As systemic steroids remain the main stay of treatment we started him on topical application of clobetasol propionate- tid for 2 weeks along with clotrimazole cream two times for 2 weeks and lignocaine viscous mouth rinse. after dermatology consultation for lesion on his glans penis he was given candid skin cream for local application. Patient was asked to completely avoid hot and spicy foods. On review after 2 weeks there was no signs of reduction and hence we started him on systemic steroids after checking his blood counts and oncologists consultation. He was started on prednisolone 10mg bd for 1month and topical application of clobetasol propionate was reduced to local application twice daily along with vitamin, iron, calcium supplements and antacids. Fluconazole was also give 100mg 1od for 2 weeks. Review was done after one month and on examination there was great reduction in the size of the ulcerations and patient was able to start with solid food, with very less burning sensation, however, the desquamation in buccal mucosa along the occlusal plane was persisting, but the lesion on ventral surface of tongue had completely subsided. Then reduction in dose of steroids to 15mg/day was done for a period of one more month along with clobetasol propionate once daily for one month. Other medications were also were continued. After 1 month the lesion in occlusal plane not subsided but wicham’s striations were visible. On subsequent review azathioprine 50mg was started along with a maintenance dose with prednisolone 10mg was continued. Topical antifungals were given along with other systemic supportive medication. On review mild ulcerations were noticed on the buccal mucosa along the occlusal plane and the dose of azathioprine was increased to 100mg which could not be tolerated by the patient. Then it was decreased to 50 mg/ day. Counts were checked weekly and patient is kept under maintenance. This case of severe ulcerative lichen planus was not responding to topical steroid and low dose of systemic steroids like the most commonly seen intraoral lichen planus. This would be probably due to the malignancy and change in the immune profile of the patient which could have activated the lesion. The concomitant use of immunosuppressant along with systemic steroids would be required for the management of these kind of severe cases of lichen planus, as patient would need to continue the treatment for a longer duration of time.

DISCUSSION

Lichen planus is an autoimmune inflammatory condition that can affect any part of oral cavity, skin, nails, etc. It usually responds to topical or systemic steroids. The temporal association of lichen planus with lymphoma in this patient suggest that neoplasm may have triggered the development of lichen planus. Though the biopsy report was suggestive of lichen planus, patient has severe stomatitis. The erosive nature caused severe sloughing of mucosa associated with stomatitis to suggest an autoimmune vesiculobullous lesions like pemphigus vulgaris, bullous pemphigoid or paraneoplastic pemphigus. Helm et al in 1994 speculated that tumor antigens may lead to production of autoimmune response leading to lichen planus in patients with neoplasia. In neoplasia induced lichenplanus mucocutaneous reaction patterns occur with diverse triggers ranging from medicaments to neoplasia. Neoplasia induced lichen planus is described as a cell mediated reaction to unknown epithelial antigens. Ours was a case of severe recalcitrant type of lichen planus which was treated with systemic steroids along with immunosuppressants like azathioprine.

![Fig 1](image1.jpg)

**PREOPERATIVE**

**POSTOPERATIVE**
REFERENCES


ABSTRACT
We report a case of Freeman-Sheldon syndrome with Congenital Vertical Talus in a 10 month-old child, which was treated by closed manipulation using a newer management protocol. Adeelan Score was used to assess the progress and outcome of treatment. The procedure involved serial manipulations by Reverse Ponseti method and casting followed by limited open realignment and fixation.

The clinical and radiological results at the end of two years were very good. The child is on an ankle foot orthosis and kept under regular follow up. The potential advantage of this method is that the structured serial manipulations stretch the soft tissues. Hence the soft tissue release can be kept to a minimum, which also helps in open realignment of the bones. Less soft tissue release also reduces scar tissue formation, thus maintaining the flexibility of the foot.

Key words: Freeman-Sheldon syndrome, Congenital Vertical Talus, Reverse Ponseti method

INTRODUCTION
Congenital vertical talus, also known as congenital convex pes valgus, is an uncommon foot deformity that is present at birth and has an estimated incidence of 1 in 10,0001. It is characterized by a fixed dorsal dislocation of the navicular on the talar head and neck. Bilateral involvement in 50 percent of cases2. The pathoanatomy of Congenital Vertical Talus has been well documented1,2. It was described first by Rocher3 in 1913 who called it “foot in piole”. The characteristic features was described by Lemi & Weissmar2 in 1939. The exact etiology is still unknown but over pull of anterior Tibial tendon in paralytic disorders and Intrauterine compression has been described as a possible cause.

The deformity may also occurs following a degree of growth arrest at 7th and 12th weeks of gestation7,8. Hamanishi4 first classified CVT in five groups and described about the concept of TAMBA (Talar axis first metatarsal base angle) & CAMBA (Calcaneal axis first metatarsal base angle). HOX gene mutation may cause Congenital Vertical Talus in many families and in Charcot Marie tooth disease5,6. However, unlike clubfoot, essentially 100% of reported vertical talus deformities have not been fully corrected with cast immobilization alone and have required major reconstructive surgery10,11.

Initially surgery was the mainstay of deformity correction and the treatment was according to age and severity of deformity. Surgical option included Open Reduction & realignment of Talonavicular & subtalar joints and a combination of several soft tissue release and Subtalar and triple arthrodesis for permanent correction of the deformity11,12,17,18.

But over the last five years the possibility and efficacy of a New method of treatment for Congenital Vertical Talus has been explored. The main components of the new methods are Reverse Ponseti cast & Minimal invasive procedure.

Freeman-Sheldon syndrome (FSS) is a rare form of the multiple congenital contracture (arthrogryposis) syndromes, also known as whistling face syndrome, characterized by dysmorphic status combining bone anomalies and joint contractures with typical facies features.21,22 FSS is part of the nosologic group of the distal arthrogryposis. The three basic abnormalities are microstomia with pouting lips, camptodactyly of the hand & talipes equinovarus/vertical talus.

CASE REPORT
A 8 months old male child presented with deformity of both feet since birth.

Antenatal history revealed that he was the 1st baby of diabetic mother born via LSCS and birth weight being 2.8kg.

Examination revealed that forefoot was in dorsiflexion and abduction whereas the hind foot was in valgus. The sole was convex with prominent medial border & antero-lateral and posterior crease.

The degree of deformity was more in left foot when compared to right foot.

A Head to Foot examination revealed that child is having Plagiocephaly, narrow palpebral fissure, high arched palate, Micrognathia, short neck posterior hair line, hands showed thumb in palm deformity & Camptodactyly of 2-5 fingers on both sides.

Upon considering the above mentioned features with CVT the diagnosis of Freeman Sheldon syndrome was entertained. The confirmation of diagnosis of Congenital Vertical Talus was done Radiographically.

Department of Orthopaedics
Congenital Vertical Talus In Freeman- Sheldon Syndrome. Treated with New Method (Reverse Ponseti method).

Fig-1 rocker bottom foot with anterolateral crease Dorsiflexion foot. Fig-2A posterior crease Fig-2B Reverse Ponseti cast.

Fig-3 - AP VIEW - TAMBA - 31 DEGREE. Fig-4 - AP VIEW – TAMBA – 43 DEGREE

Fig-5 - LAT.VIEW - TIBIOTALAR - 157 DEG. Fig-6-LAT.VIEW - TIBIOTALAR ANGLE - ANGLE 145 DEG.
To proceed further we classified the Congenital Vertical Talus According to Coleman’s classification system. We found that this case fall in Coleman’s Type 1 group.

We also scored the deformity of foot according to Adeeliar Score & it was –8 / 10.

With the above preparations treatment was started according to New method (Reverse Ponseti technique) at 10 months of age. Serial manipulations and reverse Ponseti casting was done and total of 9 casts was applied.

During the course of casting, deformity of feet was scored after each casting and it was found that there was dramatic change in clinical appearance of foot & improvement in Adeeliar score, But radiological parameters were improving slowly. Following the 9th cast the situation was analyzed both clinically & radiographically.

Clinical assessment revealed a near normal appearance of feet with anterior & lateral crease absent though posterior crease persisted. Heel valgus was present. Plantar flexion of 40 degrees was possible.

Radiographic assessment of the included angles was done as is shown in Table-2.

<table>
<thead>
<tr>
<th>ANGELS</th>
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<th>OBSERVED (AC)</th>
<th>NORMAL</th>
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<td>AP VIEW</td>
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<td>10</td>
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<td>TALUS AXIS 1ST METATARSAL AXIS ANGLE</td>
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<td>LATERAL VIEW</td>
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<tr>
<td>TAMBA(PF)</td>
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<td>64</td>
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Having met the prerequisite for surgical correction, we proceeded to the surgical correction of residual deformity which was achieved by open reduction & k wire fixation of Talonavicular joint after realigning & reducing the talus, followed by percutaneous tenotomy of Tendoachillis tendon. Five degree dorsiflexion groin to toe cast was given in immediate postop period. After 3 weeks cast was removed & reapplied in 15 degrees of Dorsiflexion, after 5 weeks k wire was removed and brace was given in 10 degrees plantar flexion and 10 degrees of adduction till walking age. Ankle foot orthosis was given in with 15 degrees plantar flexion and 15 degrees adduction. Throughout the follow up period parents are counseled as to the importance of maintaining the achieved result and being compliant. ROM exercises and foot inversion were done 2-3 times per day.
At last follow up at 18 months of age.

Child is walking with normal appearance of foot with normal range of motion of ankle & on AFO for maintenance.

Follow Up -

Fig-14 - X ray ap & lateral view - follow up at 18 months showing reduced talus & calcaneum.

Fig-15 - Braces showing 15 degree dorsiflexion & 15 degree adduction at tmt joint level.

REFERENCES


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