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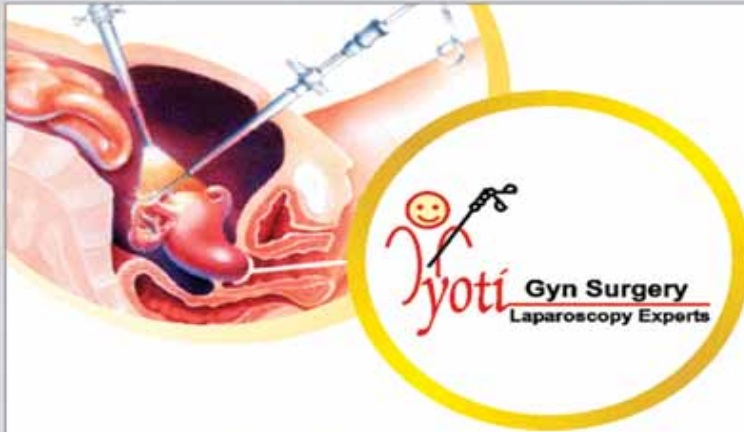
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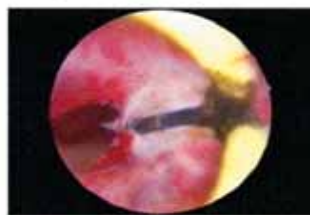
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## From The Editor's Desk.....



Dear Friends,

Let me wish a very happy new year to all. I am pleased to introduce myself as the incoming editor to BJ kines and feel honored to take up this role. BJ Kines is already indexed in Copernicus, but I wish to see BJ kines develop into a scientific journal with its balance shifting towards research communication rather than remaining a bulletin. The success of this enterprise depends on your response and I would appreciate your feedback to help achieve this goal. I would like to encourage you to submit original research notes as well as review articles, pictorial CME, case reports, short communications & letters to the editor which would comment on some important medical issue or give feedback on the articles in previous issues. We are also giving you guidelines and request that you submit your articles accordingly. Under the able guidance of our Dean, Dr B.J Shah, let us join hands and make our journal an index journal. I sincerely thank all the members of editorial board without whose help the journal could not have seen the light of the day.

**Dr. Asha N Shah**

M.D. Medicine  
Professor & Head,  
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## Taking Small Steps Towards A Big Success..

Department of Paediatrics was started in the year 1911 at the New Civil Hospital, Asarwa with provision of indoor facility of 50 beds.

Our Neonatal Intensive Care Unit was inaugurated in the year 1982 and was a dream project of Dr. A.B.Desai which was then expanded by Dr..S.N.Vani aiming at intact survival of newborn.

Over a period of years it has grown to current status just like a neonate grows into an infant with the bed capacity of 80 beds, highest among all Medical Colleges of Gujarat

It was a proud moment for Department of Paediatrics & Professor and Head of the Department, Dr. K. M. Mehariya to receive an Appreciation Award for Best Nursery amongst all Medical Colleges of Gujarat, awarded by the State Government of Gujarat (Health Department) and the honours were done by our Honorable Minister of State For Health, Shri Jaynarayan Vyas and NRHM-State Mission Director, Ms. Anju Sharma on 7<sup>th</sup> April 2012.

Our newly constructed "VATSALYA" neonatal care unit was inaugurated in the esteemed presence of our Medical Superintendent Dr. M. M. Prabhakar and our Dean Dr. B. J. Shah on 27<sup>th</sup> January 2012 and visited by Ms. Anju Sharma, Mission Director NRHM.



**New Neonatal Intensive Care Unit:**  
**"VATSALYA"** under department of Paediatrics,  
 inaugurated by Dr. M. M. Prabhakar,  
 Medical Superintendent, CHA, &  
 Dr. B. J. Shah, Dean, B. J. Medical College, Ahmedabad.

The State Government Health Fund and Health Fund granted by our medical Superintendent has enabled us to provide total free health services like higher antibiotics, immunoglobulin therapy for sepsis, surfactant therapy for respiratory distress in newborn.

Under Janani Shishu Suraksha Yojana (JSSY), an initiative of government to reduce MMR and IMR, there is a provision to provide free medication to every newborn till 28 days of life. We also provide free H.Influenza type b vaccine under this programme.

We have a separate High Risk Newborn Clinic started recently with dedicated faculty and resident staff over and above the Well Baby Clinic that takes care of postnatal babies. At this High Risk Newborn Clinic, each baby will be subjected to

1. Neuro developmental assessment
2. Retinopathy of prematurity (ROP) screening
3. Hearing Assessment

as per the discharge and follow up criteria.

A novel project of State Health Department for Neonatal screening was started for intramural babies for early detection of fatal/lethal and/or treatable inborn errors of metabolism including congenital hypothyroidism.

The departmental faculties are conducting year round EmNBC (Emergency Newborn Care) training of staff nurses and medical officers from periphery (PHC, CHC and District Hospitals) to ensure that no newborn is left unattended at birth.

To invigorate our commitment to newborn care, last year state level conference on neonatology (NEOCON 2011) was organized by our department in collaboration with IAP-GSB Ahmedabad branch and National Neonatology Forum Gujarat, which was a grand success.

We are proud to announce that we have expanded our services in paediatric super speciality branch of Neurology, Paediatric Intensive Care with the help of Paediatric Neurophysician and Paediatric Intensivist round the clock.

Apart from that our department extends its helping hands in rural areas in form of school health services-supervisory visits by faculties every year ever since the inception of the programme.

Our department is committed to public health and every year we conduct awareness programme, for diarrhoeal disease, thalassemia, breast feeding promotion, newborn care practices in community.

As is rightly said by Ben Sweetland,

**"SUCCESS IS A JOURNEY NOT A DESTINATION"**  
 Department of Paediatrics will continue its journey towards improved neonatal outcome with full dedication and countless efforts.

# Diabetic Autonomic Neuropathy

\*Asha Shah, \*\*Roopesh Singhal, \*\*Dinesh Joshi, \*\*Pallav Parikh, \*\*Mitali Desai,  
\*\*Abhinav Jain, \*\*Pinakin Patel

Diabetes has evolved to a stage where it requires no introduction to acknowledge its importance and implications. Some aspects of Diabetes however seem yet far from understood and familiar. One such condition is Diabetic Autonomic Neuropathy (DAN). DAN is an important, serious, common and yet largely under recognized complication of diabetes. DAN can affect all limbs of the autonomic nervous system and have manifestations involving all organ systems.

DAN has evaded the attention of patients and physicians alike. Patient often have minor symptoms of dizziness, light headedness that tend to be ignored both by the patient and the physician; thus DAN remains under appreciated and under treated. The clinical manifestations of DAN can range from minor discomfort to potentially life threatening situations; though in quite a few, it may still be asymptomatic.

The common features are resting tachycardia, exercise intolerance, orthostatic hypotension, constipation, diarrhoea, sudomotor dysfunction, hypoglycemic unawareness, erectile dysfunction etc.

The reported prevalence of DAN varies widely. This highlights the lack of a standardized definition and diagnostic criteria. Though the prevalence of DAN increases with duration of diabetes it has been found to present at the time of diagnosis in both type I and type II diabetes patients.

Sub Clinical DAN has been identified as early as 2 years of type I diabetes and 1 year of type II diabetes. Recently, there have been studies even describing autonomic dysfunction in patients with pre- diabetes. It can be said without doubt that the iceberg of DAN goes much deeper than previously fathomed. Numerous postulations exist explaining the occurrence of DAN (**Table 1**).

**Table : Pathogenic factors implicated in development of DAN**

Pathogenic pathway	Mechanism
Polyol metabolism	Increased accumulation of sorbitol intracellular myoinositol deficiency, reduction in the activity of protein kinase C and Na/K-ATPase
Fatty acid metabolism	Accumulation of linoleic acid and removal of linolenic acid changes in membrane properties and decreased synthesis of vasoactive substances followed by hypoperfusion of nerves
Glycation of Proteins	Non-enzymatic glycation of proteins formation of advanced glycation end products with changes to protein structure, function and immunogenicity
Endoneural ischaemia/hypoxia	Decrease of endoneural circulation local ischaemia
Oxidative stress	Increased formation of oxygen free radicals and destruction of antioxidant mechanisms tissue damage
Destruction of nerve growth factors and axonal transport	Reduction in nerve growth factors and their receptors destruction of the synthesis of neural proteins
Immunological processes	Autoimmune reactions, including inflammatory processes

Adapted from Luft.

Hyperglycemia and related metabolic insult are said to be central to most of these mechanisms. Recently though various inflammatory pathway have been implicated in

pathogenesis of DAN.

Cardiovascular autonomic dysfunction is the most studied component of DAN. CAN is associated with dysfunction of both sympathetic and parasympathetic limbs of ANS. The reported prevalence of CAN varies from 2.5% to 90%; it is about 20% in asymptomatic diabetics. Of particular interest is the association of CAN

\* Professor & Head

\*\* Resident, Department of Medicine,  
B. J. Medical College, Ahmedabad.

with silent MI and sudden cardiac death. Reduced heart rate variability (HRV) is one of the earliest features of CAN. Meta-analyses of 12 studies demonstrated that reduced cardiovascular autonomic function (measured by reduced HRV) had relative risk 1.96 times for silent myocardial ischemia and mortality.

It is known that parasympathetic component is affected earlier than sympathetic counterpart. It is the withdrawal of resting parasympathetic tone that is said to be accountable of resting tachycardia. Orthostatic hypotension is largely accounted by loss of sympathetic response to sudden postural changes. Affection of the sympathetic part also brings down the heart rate a bit but with profound loss of the heart ability to vary pulse rate in response to physiological needs. Of particular mention is

the decreased sympathetic response to hypoglycemia leading to hypoglycemic unawareness; the problem is only exaggerated by increased frequency of such attack due erratic sugar levels in DAN or the so called “brittle diabetes”. Heart response to Valsalva maneuver involves both the components depending upon the stage of maneuver. It is this dynamic balance between the sympathetic and parasympathetic tone that is studied by means of HRV and it is loss of these oscillations that suggest autonomic dysfunction.

Diagnosis of CAN is to be made by a group of clinical tests rather than a single test alone; of particular significance are tests using HRV which have reliability and reproducibility needed for studies.

Diagnostic tests for the two components are given in **Table 2**.

**Table : Diagnostic Test for Autonomic Dysfunction**

Parasympathetic	Sympathetic
<ul style="list-style-type: none"> <li>• Resting heart rate</li> <li>• Beat to beat variation with breathing (E:I ratio)</li> <li>• 30:15 heart rate ratio with standing</li> <li>• Valsalva ratio</li> <li>• Spectral analysis of heart rate variation (high frequency power)</li> </ul>	<ul style="list-style-type: none"> <li>• Resting heart rate</li> <li>• Spectral analysis of heart rate variation (very-low frequency power)</li> <li>• Orthostasis blood pressure</li> <li>• Hand grip blood pressure</li> <li>• Cold pressor response</li> <li>• Sympathetic skin galvanic response</li> <li>• Sudometry</li> <li>• Cutaneous blood flow</li> </ul>

Adapted from Vinik and Ziegler

Methods for analysis of HRV can be described as time domain based measures (which involve using RR intervals mean, differences, standard deviation over etc) or spectral analysis or frequency domain based. Spectral analysis breaks down the RR interval variation as function of set of sine waves using methods like Fourier transformation; frequency based group of these waves are seen to associate with specific autonomic component. Parasympathetic discharge corresponds to high frequency waves (0.15 – 0.4 Hz) and Sympathetic to very

low frequency waves (<0.04 Hz); low frequency (0.04 – 0.15 Hz) is seen to be influenced by both components. The ratio of very low frequency to high frequency can then be used to quantify autonomic dysfunction. The main advantage with spectral analysis is it is much more objective and requires lesser patient cooperation. Ansiscopes a simple portable noninvasive device uses spectral analysis of 512 RR intervals to quantify autonomic dysfunction.

Based on the obtained percentage patients can be classified as having normal, early, late, advanced, most advanced autonomic dysfunction (**Table 3**).

**Table 3 : Classification of Autonomic Dysfunction Measured by ANSscope**

Category	% Quantification	Implications
Healthy	0 – 20 %	No autonomic dysfunction
Early	21 – 40 %	No organs affected yet, only functional disorders occur
Late	41 – 60 %	Some probability that organs may be affected. Constitutes rare chance of returning to previous groups
Advanced	61 – 80 %	High probability of organs being affected
Most Advanced	81 – 100 %	Complete failure of one or both subsystems of ANS.



More recently, dynamic pupillometry has been described measuring the latency of papillary reaction to flashes of light for even earlier detection of autonomic dysfunction.

QTc interval is known to increase in DAN; it can be taken as a surrogate marker for HRV. We evaluated the simple pulse measurement and QTc intervals against the newer sophisticated methods of assessment; and found a significant association between QTc prolongation and DAN occurrence and grade. QTc interval prolongation is known to increase the risk of developing serious ventricular arrhythmias and sudden cardiac death; and the occurrence of such events may increase in presence of CAN. In all patients with features of advanced autonomic neuropathy, 12 lead ECG should always be obtained and looked for QTc prolongation. Similarly in all diabetic

patients with QTc  $\geq 440$ ms, other features of advanced autonomic neuropathy should be searched and treated.

It is important to remember that the response to treatment will depend upon how early the treatment is initiated. Thus early treatment may not only halt the progression but also partly reverse the autonomic dysfunction. Various drugs have been implicated in treatment of DAN are ACE – inhibitors, ARBs, cardioselective  $\beta$  blockers, antioxidants, aldose reductase inhibitors etc but of most paramount importance is tight glycemic control. We must realize with established neuropathy tight glycemic control becomes more risky and it is best that DAN be prevented by proper glycemic control in the first place.

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# Exercise awareness among medical students of B J Medical College, Ahmadabad.

Jyotsna Tabiyar\*, Puja Chaudhari\*, Mitali Leuva\*\*, Tejas Shah\*, Paresh Prajapati\*, MK Lala\*\*\*

## ABSTRACT

**Introduction:** Exercise provides many physiological and psychological benefits. Available experience and scientific evidence show that regular exercise provides physical, social, mental health benefits. Despite many benefits of exercise behavior, physical inactivity is a global health issue and there is evidence to indicate that the exercise training of medical students is inadequate in both quality and quantity. **Material and method:** It was a cross sectional study conducted on under graduate medical students of B.J.Medical, Ahmedabad. 200 under graduate medical students were selected on random basis. On predesigned exercise behavior questionnaire perform, details were noted. **Results:** 53% of students had knowledge about exercise. 70% of medical students had no knowledge about side effect of exercise. There were 42% medical students suffering from pre existing conditions. 82% of students had not consulted any medical person for what kind of exercise should be done. **Conclusion:** Even though, there were medical students, they had not knowledge about consulting a medical person for what kind of exercise should be done, they want to do. Thus by studying exercise awareness among medical students and advising them for exercise we can prevent occurrence of NCD risk factors among them and psychological problems could be decreased.

## INTRODUCTION

Exercise is a global health issue. It provides many psychological and physiological benefits<sup>1</sup>. It is necessary for everyone at all stages of life<sup>2</sup>. But inactivity is common in all age groups especially among young generation. Regular exercise contributes to a healthy, independent life style and greatly improves the quality of life. According to scientists, exercise therapy is successful because of the balance created in the nervous and endocrine systems which directly influences all the other systems and organs of body<sup>3</sup>. Exercise acts both as curative and preventive therapy. Through the practice of

exercise we become aware of the interconnectedness between our emotional, mental and physical levels<sup>3</sup>. Regular practice of exercise can help blood pressure, digestive disorders, arthritis, arteriosclerosis, chronic fatigue, asthma, varicose veins and heart conditions. Laboratory tests have proved the exercise increased abilities. Overall physical inactivity was estimated to cause more than 2 million deaths<sup>4</sup>. The minimum amount of physical activity required for prevention of disease is at least 30 minutes of moderate activity every day<sup>5,6</sup>. Habit of exercise is not very popular among young generation today and also Outdoor games are not popular. Younger generation usually sit on computer & work. With this handover this study has been done to know knowledge & habit of exercise among undergraduate.

## MATERIAL AND METHODOLOGY

The present study is conducted in B J Medical College; Ahmadabad .Ahmadabad city is located on the bank of river Sabarmati. Ahmadabad is the largest city of Gujarat & 6<sup>th</sup> largest city of India.

**Study design :** Cross-sectional study

**Study area :** B J Medical College, Ahmadabad.

**Study duration :** December 2011 to January 2012

**Data collection :** a pre-designed and pre-tested Performa was used to collect baseline data .Informed consent was taken before the initiation of survey and information is collected regarding exercise awareness among undergraduate medical students of B J Medical College, Ahmadabad.

**Statistical analysis :** analysis was done in epi - info version 3.5.1

## RESULTS

**Table I. Distribution of students according to their exercise habit**

Habit of exercise	No of Students (Percentage)
Yes	106(53)
No	94(47)
Total	200(100)

\* P.G. students,

\*\* Tutor,

\*\*\* Associate professor,

Department of Community Medicine,  
B. J. Medical College, Ahmedabad.

**Table II. Knowledge of adverse effects of exercise**

Knowledge on adverse effect of exercise	No of Students (Percentage)
Yes	62(31)
No	138(69)
Total	200(100)

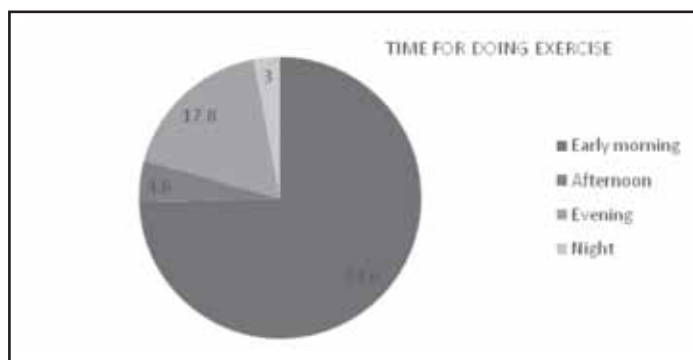
**Table III. Distribution of Medical students on consultation to any Physician**

have you ever consulted any physician/physiotherapists/trainer/professional for kind of exercise	Frequency (percentage)
NO	160(80)
Yes	40(20)
Total	200(100)

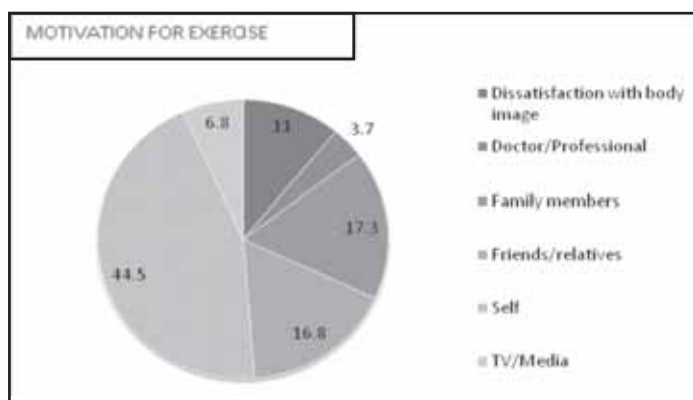
**Table IV Reason for going to gym**

if you go to gym then why	Frequency	Percent
fitness	85	86.7%
make friends	4	4.1%
pass time	3	3.1%
relax	6	6.1%
Total	98	100.0%

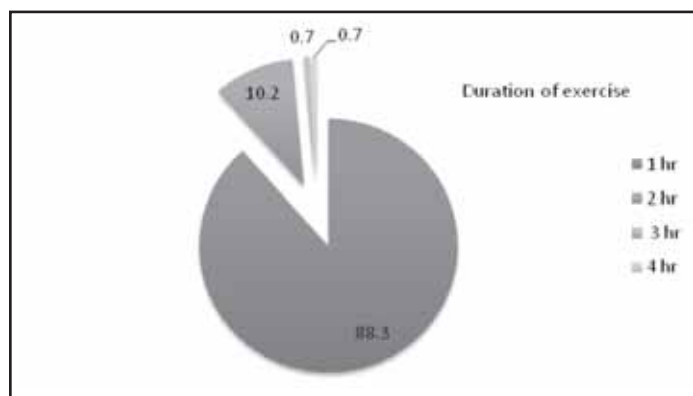
**Figure I**



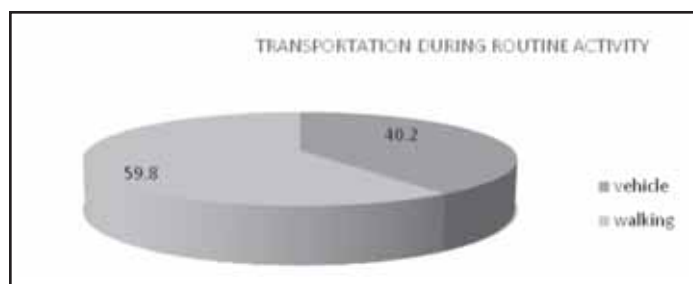
**Figure II**



**Figure III**



**Figure IV**



## Discussion

53% of medical students had knowledge about exercise. Most of medical students had no knowledge about the side effect of exercise. 21% medical students were suffering from pre existing conditions. Most common pre-existing condition was lack of concentration. Most of students had not consulted any medical person for what kind of exercise should be done. 59.8% of them preferred to go by walking for routine activity. 44.5% of medical students were self motivated for starting exercise. 74.6% preferred to do exercise early morning. Even though they were medical students, they did not have knowledge about consulting a medical person for what kind of exercise should be done. Thus by studying exercise awareness among medical students and advising them for exercise we can prevent occurrence of various health related problems among them. Creating awareness about doing exercise among medical students and also about psychological and mental benefits of exercise. Thus we can increase the number of students preferring physical activity. There is a need to carry health survey of undergraduate students annually so that pre existing health condition among them can be detected and required exercise habit can be advised. Facility may be provided at medical colleges for outdoor game include. In the era of NCD there is needed to include exercise related chapter in undergraduate curriculum.

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# Knowledge, Attitude And Practice Regarding Dietary Habits In Medical Students

Tejas Shah\*, Paresh Prajapati\*, Jyotsna Tabiyar\*, Atul Trivedi\*\*, Mitesh Patel\*\*

## ABSTRACT

Nutrition interferes significantly with every field of medicine<sup>1</sup>. To exclude or ineffectively approach the nutrition correlates of disease is to risk an increase in morbidity, mortality, from many potentially preventable diseases and healthcare costs of hospitalised patients<sup>2</sup>. most curricula do not provide identifiable period of training in nutrition and many medical schools lack adequate curriculum on these topics<sup>3</sup>. The solution to above dilemma is to raise awareness through training of medical students regarding importance of concepts of nutrition.

**Key words :** Nutritional knowledge, eating habits, practice

## Introduction

Nutrition is an important component in the treatment of acute and chronic diseases and is cornerstone in strategies for disease prevention and health promotion. Nutrition also plays an important role in the etiology of many diseases. Despite the acknowledged importance of nutrition, there is evidence to indicate that the nutrition training of medical students is inadequate in both quality and quantity<sup>1</sup>. The study aimed to know the dietary habits of medical students, assess their knowledge on nutrition and to assess their confidence in addressing the dietary issues of patients.

## Methodology:

It was a cross sectional study conducted on final year under graduate medical students of B. J. Medical College, Ahmedabad. The sampling was purposive and total of 104 students participated in the study voluntarily. A pretested and predesigned questionnaire was used to assess their nutrition knowledge, their eating habits and overall perception regarding importance of healthy eating habits. The questionnaire contained multiple choice questions. Habit of eating breakfast, consuming

milk, fruits and green leafy vegetables was assessed. The replies classified as frequent were “consumes daily” or “alternate day” and as infrequent were “once in a while” or “don't consume”. More frequent habit of eating breakfast, milk, fruits and greens and less frequent habit of consuming soft drinks and snacks were considered healthy. The level of knowledge in nutrition was assessed by questions on sources of carbohydrates, fats, proteins, vitamins and minerals. The replies were categorised in “more knowledgeable” when score was >75<sup>th</sup> percentile and “less knowledgeable” when score was <75<sup>th</sup> percentile. For diet, the students rated their confidence using questionnaire in ability to assess patients diet, help patient change diet and make recommendations to them.

## Observations

A total of 104 students participated of which males were higher(56%) compared to females(44%). Majority of students are having healthy eating habits(table 1). Overall, 90% of students are aware regarding importance of The level of knowledge among students shows that 45% are more knowledgeable and 55% are less knowledgeable(table 2,3). There is no association between there healthy eating habits and more knowledge( $p>0.340$ ). Only 45% of students were confident in assessing the diet of patients. The present study shows no association between increase in level of knowledge and confidence level of students in addressing dietary issues( $p>0.3$ ). Majority of students(81%) responded that medical curriculum is inadequate to teach nutrition(table 5). Most students(56%) were of the opinion to have separate department for teaching nutrition(table 6).

## Results

### I. Frequency Breakfast, Milk, Fruits, Green Leafy Veg Consumption

	MALE	FEMALE
YES	52 (89.7%)	37 (80.4%)
NO	6 (10.3%)	9 (19.6%)

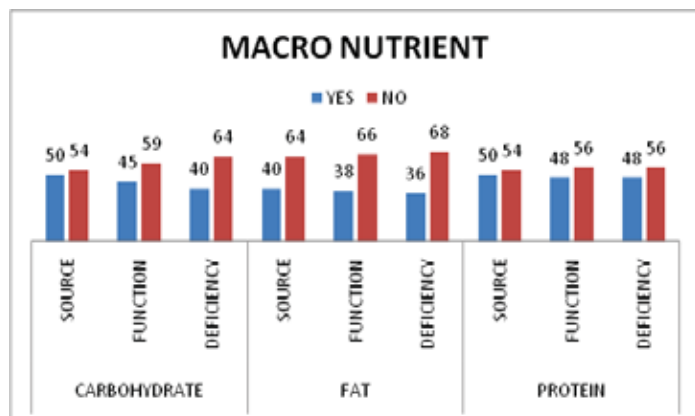
**Majority of Students are having Healthy eating habits [80.4% & 89.7%]**

\* Resident,

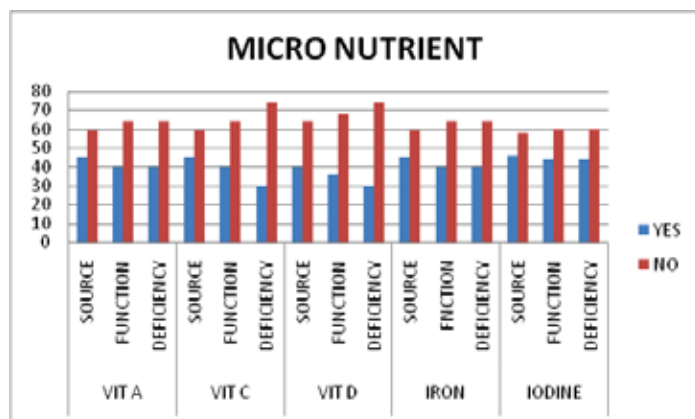
\*\* Assistant Professor

Department of Community Medicine,  
B.J.M.C, Ahmedabad

## II. Knowledge Regarding Diet-Macronutrient [Source, Function & Deficiency] :



## III. Knowledge Regarding Diet-Micronutrient [Source, Function & Deficiency] :



Both Tables show Inadequate knowledge of students regarding nutritive substances

## III. Opinion regarding importance of healthy eating habits in daily life :

	Male	Female
Yes	51 (89%)	41 (91%)
No	4 (7%)	3 (5%)
Don't know	3 (5%)	2 (4%)

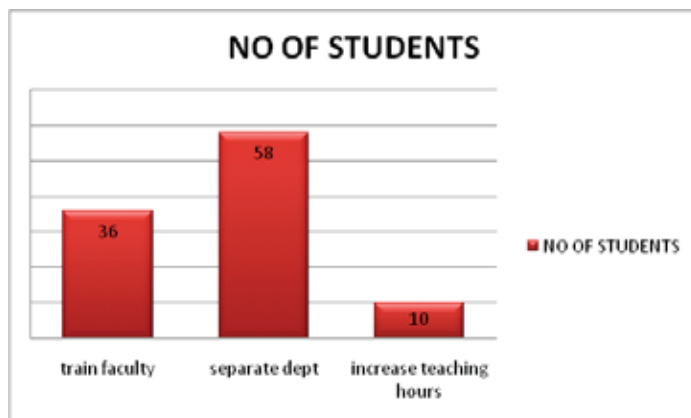
Thus, 90% of students are aware about importance of healthy eating habits

## IV. Response of medical students regarding medical curriculum able to teach nutrition :

	Male	Female
Not enough	47 (80%)	38 (83%)
Just enough	8 (14%)	5 (11%)
enough	3 (6%)	3 (6%)

Thus, majority of students [81%] responded that current medical curriculum inadequate to teach nutrition

## V. Suggestions to improve learning in nutrition :



Majority of students [55.76%] were of the opinion to have separate dept for teaching nutrition

## Discussion

The knowledge in nutrition was assessed among medical students and it was observed that 55% were less knowledgeable and 45% were more knowledgeable. Ammermann et al<sup>4</sup> studied the relationship among dietary knowledge, attitude and behaviour to identify educational needs of entering medical students and found that more favourable score were associated with healthier eating habits, but greater knowledge was not. Similarly, 90% of students in this study group have healthy eating habits and there is no association between their healthy eating habits and more knowledge ( $p > 0.340$ ). This study demonstrates 62% of students were not confident to assess diet of patients and recommend change in it. In a similar study done by Conroy et al<sup>3</sup> among medical students at Harvard Medical School revealed the baseline confidence of students in ability to assess and counsel about diet is low. Conroy et al<sup>3</sup> showed that confidence of students improved after nutrition course ( $p < 0.001$ ). This proves that more effective nutrition training course is required to improve confidence of students. Winick<sup>5</sup> identified the barriers in nutrition education and found limitation in teaching time and faculty members for teaching nutrition. This study concludes that comprehensive nutrition curriculum is needed including training of faculty or creation of separate nutrition department. The study results intend to stimulate active consideration of proper role of nutrition learning in medical education.

## Results & Conclusion :

Overall 90% of medical students were aware regarding importance of healthy eating habits and were practicing it in their daily life. However, only 40% of students were knowledgeable about nutrition. There was no statistical association between their healthy eating habits and more knowledge regarding nutrition ( $p>0.3$ ). Most students (80%) were of the opinion that adequate nutritional knowledge was not imparted to them and expressed keen desire to study nutrition as separate topic both academically and clinically. 60% of students said that faculty of different clinical departments should be trained in nutrition. The study results intend to stimulate active consideration of proper role of nutrition learning in medical education.

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# A Rare Case Of Penile Fracture Managed By Surgical Exploration And Repair

Shrenik Shah\*\*\*, Ketan Shukla\*\*, Bodhraj Pasi\*, Abhinav Kumar\*, Rakesh Pandey\*,  
Prakash Chaudhary\*, Bharat Chheta\*

## ABSTRACT

The diagnosis of “penile fracture” specifically refers to a rupture of the corpus cavernosum induced by blunt trauma to the erect penis. Prompt surgical exploration and corporal repair is the most efficacious therapy. We present a case report of a male presented with penile fracture. Urgent exploration & repair of penile fracture was done on the basis of his clinical presentation. With our experience, we suggest that prompt surgical management is ideal in cases of penile fracture.

**KEYWORDS-** Penile fracture

## Introduction

Penile fracture typically occurs when the engorged penile corpora are forced to buckle and “pop” under the pressure of a blunt sexual trauma. Here we present a rare case of penile fracture managed by surgical exploration and repair.

## Case Report

A 43-year-old, healthy male presented to the Urology Department with acute scrotal pain and penile swelling lasting 48 hours after traumatic intercourse. The injury occurred when his penis slipped from the vagina and was forcefully thrust against his partner's perineum. Upon injury, he heard a “pop” from his penis, and rapidly experienced acute penile pain and detumescence. He developed swelling of his scrotum and significant pain in his penoscrotal region. He gave history of slight painless hematuria which was not associated with clots.

On examination his penis was symmetrically swollen and there was blood at the meatus. The penile shaft lacked a palpable defect & the rolling sign was absent. The penis & scrotum were markedly ecchymotic and tender. His urinalysis was suggestive of microscopic hematuria.

Ultrasound showed right corpora cavernosal soft tissue replaced by hypoechoic collection with few septations & internal echoes, surrounding soft tissue appeared bulky and edematous with few foci of calcification noted in left corpora cavernosa. Penile urethra appeared normal.

Fluid with internal echoes was seen in left scrotal sac suggestive of encysted hydrocele. On the basis of his clinical presentation, the patient was taken to the operating room urgently for exploration and repair of the acute penile fracture.

## PRE OP PHOTO



## Discussion

### Incidence and Pathophysiology

The diagnosis of “penile fracture” specifically refers to a rupture of the corpus cavernosum induced by blunt trauma to the erect penis. The tunica of the erect penis thins to approximately 0.25mm on expansion, and the firmly engorged corpora under the strain of buckling can generate pressures in excess of 1500 mm Hg and exceed the limit of the thinned tunica.<sup>1</sup>

The first case of a penile fracture was described in the literature in 1924.<sup>2</sup> The largest single series to date describes 172 cases over 9 years in a single province of Iran.<sup>3,4</sup>

### Clinical Presentation

Penile fractures are commonly diagnosed from their stereotypical clinical presentation.<sup>5</sup> Patients commonly report hearing a “pop” or cracking sound from the erect penis at the moment of injury.<sup>6</sup> Detumescence occurs rapidly, and acute swelling, pain, and penile deformity follow. The pain can vary from minimal to severe and is not proportional to the degree of injury.

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The gross appearance of a fractured penis is often summarized as an “eggplant deformity”, which refers to the combination of localized penile swelling, discoloration, and deviation towards the opposite side of the fracture.<sup>6</sup> Manual examination of the penis can often detect the site of the corporal tear by palpation of the overlying hematoma. The “rolling sign” is used to describe a firm, immobile hematoma, which is palpable as the penile skin is rolled over it.

Retrograde Urethrography and Cavernosography are indicated in suspected penile fracture that presents with voiding difficulty, hematuria, or blood at the meatus. Ultrasonography use is limited. Magnetic resonance imaging has been shown to be extremely accurate in diagnosing and localizing corporal injuries.

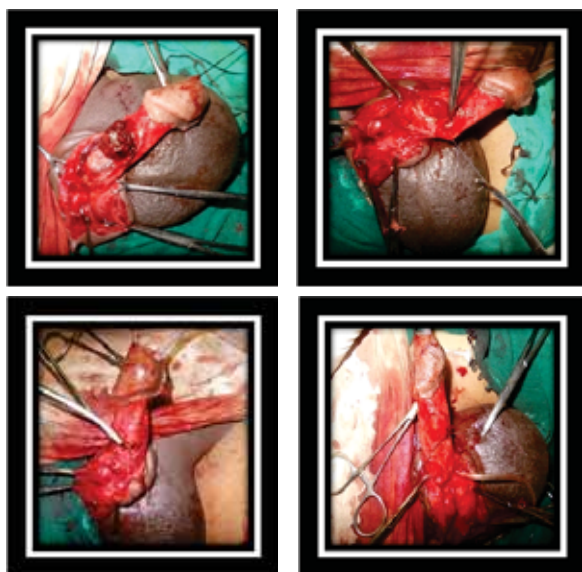
### Nonoperative Treatment of Penile Fractures

Ice packs, Foley catheterization and anti-inflammatory medicines were initially regarded as the standard of care for penile fractures, and the condition was associated with a relatively high morbidity rate. The long-term complication rates remained approximately 30% or higher.<sup>5</sup>

### Surgical Technique

Proper surgical repair of penile fractures requires evacuation of the hematoma, identification of the tunica injury, local corpora debridement, closure of the tunica lacerations, and ligation of any disrupted vasculature<sup>5</sup>. We use and recommend a distal circumferential degloving incision, as in addition to being the most cosmetic incision, it readily allows exposure to the entire tunica bilaterally, facilitating diagnosis and repair of coexisting urethral and contralateral injuries.

### INTRA OP PHOTOS



### POST OP PHOTOS



Post Operative Day 2

Post Operative Day 4

### Long-Term Sequelae of Penile Fracture

Although surgery has been shown to reduce the incidence of penile fracture complications (6% to 25%), the reported long-term sequelae after penile fracture repair include: penile deviation, painful intercourse, painful erection, erectile dysfunction, priapism, skin necrosis, arteriovenous fistula, urethrocavernous fistula, and urethral stricture.<sup>3</sup>

### Conclusion

The diagnosis of penile fracture is mostly a clinical one. Prompt surgical exploration and repair are advocated in almost all cases. Most commonly, the rupture occurs on the lateral side of the proximal corpora, but it can occur anywhere along the corpora and produce a variety of swelling patterns. Immediate surgery reduces long-term complications; posttraumatic penile curvature remains the most common long-term complaint.

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# Management Of Extra-Adrenal Paraganglioma Encasing Aorta With En Bloc Aortic Resection And Interposition Grafting

Shrenik J. Shah\*\*\*, Ketan Shukla\*\*, Abhinav Kumar\*, Bodhraj Pasi\*, Prakash Chaudhary\*, Rakesh Pandey\*, Bharat Chheta\*

## Abstract

Extra-adrenal paraganglioma is a very rare entity. Early diagnosis and surgical excision are integral part of treatment of paraganglioma. We present a case report of a male with hormonally active extra-adrenal paraganglioma that was resected laparoscopically converted to open with en bloc resection of aorta with interposition aortic grafting. Laparoscopic approach provides excellent exposure with magnification and allows proper identification of the tumour and its relation to surrounding structures; complete resection of tumour was achieved with adequate vascular control. With our experience, we suggest surgical resection with interposition aortic graft can be performed successfully in rare cases of extra-adrenal paraganglioma encasing aorta.

**Key Words** - Extra-adrenal paraganglioma

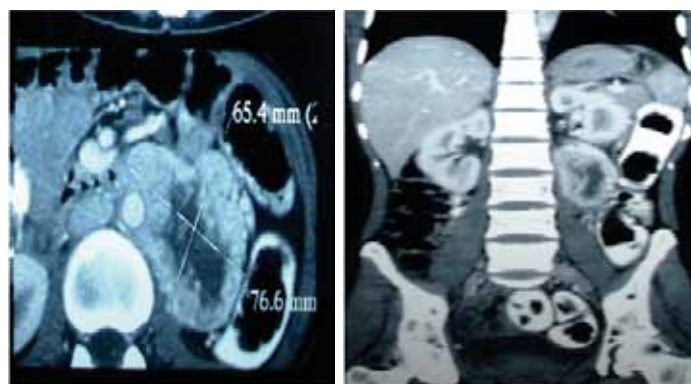
## Introduction

Paraganglioma is a rare tumour of chromaffin cells, which is frequently considered in the evaluation of hypertension. Surgery for paraganglioma differs from that of other tumours owing to the potential release of catecholamines, which may lead to severe intraoperative haemodynamic changes.

## 1. Case Report

A 28 year-old male presented with headache, palpitation and sweating for 12-month duration. His general and systemic examination was normal, except blood pressure which was 170/100 mm of Hg.

Routine blood biochemistry was normal. Computerised tomography (CT) scan of abdomen with contrast suggested –left retroperitoneal paravertebral mass lesion (heterogeneously enhancing lesion with necrosis, 92 x 76 x 65 mm size) with partial encasement of adjacent aorta - possibility of malignant neurogenic tumour.



(FIGURE-1-CT SCAN SHOWING MASS LESION)

His urinary VMA was normal (8.6 mg/24 hr). Serum levels of cortisol, testosterone and dehydroepiandrosterone (DHEA) were normal.

Laparoscopic converted to open transperitoneal resection of mass was performed under general anaesthesia. Patient was placed in the right lateral decubitus position. A 10-mm infra-umbilical port was placed using the closed technique. After establishing pneumoperitoneum, two additional 10-mm ports were introduced lateral to rectus in the left lower abdominal quadrant at the level of the anterior superior iliac spine, and the midline, midway between the xiphoid and umbilicus. Transmesenteric window was made with hook cautery. Careful dissection around the tumour at its medial border & lower pole was done. Tumour was found to be densely adherent to aorta so decision of conversion to open was taken. Tumour mass and left kidney was identified separately. Dissection around the tumour was started & all multiple feeding vessels were ligated. Vascular clamp over infra renal aorta above and below the tumour was applied and time was noted. Tumour was excised with segment of infra renal aorta. 14 no. Dacron vascular aortic graft was placed and end to end anastomosis was done with Prolene 5-0 RB continuous sutures.



(FIGURE- 2- INTERPOSITION AORTIC GRAFT)

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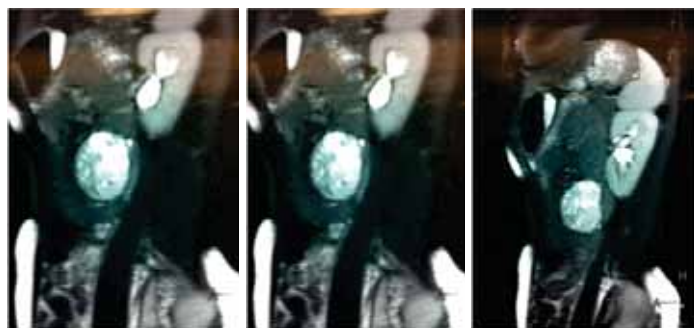
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Operative time was 220 minutes. Patient was kept for 24 hours electively in ICU for BP monitoring. Postoperative recovery was uneventful. Patient was started on liquid diet after 24 hours and soft diet after 36 hours and discharged after 72 hours. Histopathology report suggested Paraganglioma – extraadrenal with vascular invasion.

## 2 Another Rare Case Report Of Lt Para-Aortic Paraganglioma

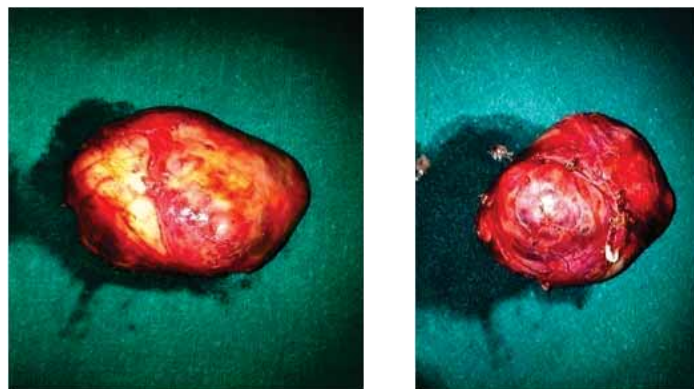
A 21 year old female patient presented with complaints of palpitations and headache since 1 month. Her general and systemic examination was normal, except BP which was 210/110 mm of Hg. Her Renal Function Tests, 24 hourly urinary VMA and Metanephrines were normal. MRI Abdomen/ pelvis suggested of left extra adrenal paraganglioma.



(FIGURE-3 MRI ABDOMEN)

Preoperatively BP was stabilised by Phenoxybenzamine 20 mg TDS and Surgery was performed by the laparoscopic route. Intra operative BP was 210/120mm Hg which was normalised after ligation of vessel to 140/84 mm Hg. Total operating time was 180min and blood loss about 80 ml. No transfusion was needed.

Oral sips were started and patient mobilised on 1<sup>st</sup> post operative day. Drain and per urethral catheter were removed on the 1<sup>st</sup> post operative day. Since patient was normotensive so no anti hypertensive were given. Discharged on 3<sup>rd</sup> post operative day.



(FIGURE-4 SPECIMEN OF PARAGANGLIOMA)

## Discussion

Paragangliomas and extra-adrenal paragangliomas are rare tumors arising from neural crest tissue that develops into sympathetic and parasympathetic paraganglia throughout the body.

## Diagnostics

The diagnosis of paraganglioma is usually suspected by the presence of an adrenal mass or a workup. Biochemical testing is done to document excess catecholamine secretion.

## Biochemical testing

### 24-hour urine collection

A 24-hour urine collection for catecholamines (e.g., epinephrine, norepinephrine, and dopamine) and fractionated metanephrines (e.g., metanephrine and normetanephrine) has a relatively low sensitivity (77%–90%) but a high specificity (98). The specificity of plasma-free fractionated metanephrines is 82% in patients tested for sporadic paraganglioma versus 96% in patients tested for hereditary paraganglioma.<sup>1,2</sup>

### Plasma-free fractionated metanephrines

Patients with symptomatic paraganglioma almost always have increases in catecholamines or metanephrines two to three times higher than the upper limits of reference ranges.<sup>3</sup>

## Imaging studies

Computed tomography (CT) imaging or magnetic resonance imaging (MRI) of the abdomen and pelvis (at least through the level of the aortic bifurcation) are the most commonly used methods for localization.<sup>4</sup>

I123-metaiodobenzylguanidine (MIBG) scintigraphy coupled with CT imaging provides anatomic and functional information with good sensitivity (80%–90%) and specificity (95%–100%).<sup>4</sup> I131-MIBG can be used in the same way, but the image quality is not as high as with I123-MIBG.<sup>5</sup>

## Conclusion

With our experience, we suggest surgical resection with interposition aortic graft can be performed successfully in rare cases of extra-adrenal paraganglioma encasing aorta. Proper patient preparation and monitoring are critical for success.



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### Answer for Quiz !!

#### Diagnosis: Congenital (hereditary) long QT syndrome.

The ECG demonstrates sinus rhythm with a very prolonged QT interval of 0.6 second. Note the broad T waves with notching (or possibly U waves) in the precordial leads. This characteristic may identify patients with long QT syndrome at increased risk for torsade de pointes and syncope and sudden death. Patients at high risk of recurrent syncope or sudden death are usually considered for implantable cardioverter defibrillator therapy along with beta-blockade. Over a dozen different "channelopathies" have been identified in the pathogenesis of congenital long QT syndrome.

T wave morphology can be helpful in determining the type of congenital LQTS. Multiple medications can cause acquired LQTS also increasing the risk of torsade de pointes and sudden death.

Visually, one should use the longest QT (often best seen in chest leads), and not restrict measurement to lead II or any single other lead. A number of rate-correction algorithms have been proposed; none has emerged as a consensus method.



# Anaphylactic reaction to Intravenous Radiocontrast agent

\*Nilima Shah, \*\*Shruti Sangani, \*\*\*Samira Parikh, \*\*\*\*Kapil Chahar

## Abstract

Radiocontrast agents are medical contrast medium used to improve the visibility of internal bodily structures in an X-ray based imaging techniques such as computed tomography (CT) or radiography. The commonly used radiocontrast agents are usually iodine or barium compounds. Modern intravenous contrast agents are usually based on iodine. Fatalities have been reported following the administration of iodine-containing contrast agents. The incidence of death is reported to range from one in 10,000 (0.01 percent) to less than one in 100,000 (0.001 percent). Here we report a case of anaphylactic reaction to radiocontrast agent, its diagnosis and management.

**Key Words :** Anaphylaxis, Intravenous pyelography (IVP), Diatrizoate meglumine, Diatrizoate sodium

## Introduction

Anaphylaxis is a severe systemic hypersensitivity reaction characterized by multisystem involvement, which may include hypotension or airway compromise. [1] It is rapid in onset and requires immediate diagnosis and treatment as it can be fatal. It represents the most dramatic and severe form of immediate hypersensitivity. This terminology now refers to both IgE and non IgE reactions.

Food, medications, insect stings, and allergen immunotherapy injections are the most common provoking factors for anaphylaxis, but any agent capable of producing sudden degranulation of mast cells or basophils can induce anaphylaxis. In India, antibiotics, radiocontrast and anaesthetic agents are considered to be the major causes for anaphylaxis. Moreover, blood products, insulin and growth hormones have also been reported to cause anaphylaxis. [2] The life time individual risk of anaphylaxis is presumed to be 1% to 3%, with a mortality rate of 1%. [3]

Anaphylaxis caused by diagnostic and therapeutic interventions is nearly unavoidable in medical practice and occurs in a variety of scenarios. Here we report a case of anaphylaxis following intravenous pyelography (IVP)

performed using ionic dye containing diatrizoate meglumine and diatrizoate sodium.

## Case Report

A 30 year male patient, diagnosed to have right sided renal calculus, was undergoing IVP procedure with an ionic dye containing diatrizoate meglumine and diatrizoate sodium. After taking consent and baseline vital parameters which were within normal limits, test dose (2 ml) of the dye was given intravenously and the patient was observed for 20 minutes. No sign of any adverse reaction was observed and hence the full dose (40 ml) was given intravenously. After 5 minutes the patient developed vomiting and palpitation. Inj. pheniramine maleate (22.75 mg) was given intravenously stat in the procedure room and the patient was shifted to emergency room (ER) immediately.

On arrival at the ER, patient was gasping and pulse was not palpable. He was intubated with 8.5 mm portex cuffed tube and intermittent positive pressure ventilation (IPPV) was started with 100% oxygen. A multipara monitor was attached to monitor other physiological parameters. Patient had bradycardia and hypotension. An anaphylactic reaction to the radiocontrast dye was suspected and inj. epinephrine (0.1mg) IV was given as 1:1,00,000 dilution followed by inj. hydrocortisone (300 mg) IV.

The patient's ECG showed ventricular fibrillation and cardiopulmonary resuscitation was started. DC shock with 360 joules was given twice at the interval of 5 mins. The rhythm was converted to ventricular tachycardia. Chest compressions and ventilation were continued between the shocks as per CPR guidelines 2010. After the DC shock, chemical cardioversion was done with inj. amiodarone 300 mg IV stat. Normal sinus rhythm was regained but the patient developed generalized tonic clonic convulsions. Inj. midazolam 2 mg IV was administered immediately followed by inj. phenytoin sodium 1000 mg (diluted in 100 ml normal saline) given over half an hour.

Once the patient's vital parameters were stable, he was shifted to ICU for ventilatory support and further management. The patient was observed in ICU for two days and treated with vasopressors and mechanical ventilation which was gradually weaned off. The patient was shifted to general ward, and discharged without any systemic or local complication.

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## Discussion

Modern intravenous contrast agents are typically iodine based. They are either organic (non-ionic) or ionic compound. The latter were developed first and are still

used widely. Ionic contrast media usually, have higher osmolality and more side-effects.

The commonly used iodinated contrast agents are described below:

Compound	Name	Type	Iodine Content	Osmolality	
Ionic	Diatrizoate (Hypaque 50)	Monomer	300 mgI/ml	1550	High
Ionic	Metrizoate (Isopaque 370)	Monomer	370 mgI/ml	2100	High
Ionic	Ioxaglate (Hexabrix)	Dimer	320 mgI/ml	580	Low

Adverse reactions to intravenous contrast media (ICM) are classified as idiosyncratic and nonidiosyncratic. Idiosyncratic reactions typically begin within 20 minutes of the ICM injection, independent of the administered dose. A severe idiosyncratic reaction can occur after an injection of less than 1 mL of a contrast agent. Idiosyncratic reactions are subdivided into minor, intermediate and severe. Severe reactions, requiring emergency measures are characterized by peripheral vasodilatation with resultant hypotension and reflex tachycardia, dyspnea, agitation, confusion, convulsions, and cyanosis progressing to unconsciousness. The histamine-liberating effect of these compounds may induce an allergic reaction which may range in severity from rhinitis or angioneurotic edema to laryngeal or bronchial spasm or anaphylactoid shock. Rarely disseminated intravascular coagulation resulting in death have also been reported. Anaphylaxis typically follow a uniphasic course, however, 20% will be biphasic in nature. The second phase usually occurs after an asymptomatic period of 1–8 hours, but there may be a 24-hour delay. Protracted anaphylaxis may persist beyond 24 hours.

Reactions to ICM have the same manifestations as anaphylactic reactions, these are not true hypersensitivity reactions as IgE antibodies are not involved. In addition, previous sensitization is not required, nor do these reactions consistently recur in a given patient. For these reasons, idiosyncratic reactions to ICM are often called anaphylactoid reactions too.

The anaphylactic reaction in our patient was probably due to radiocontrast dye as he developed signs of anaphylaxis a few minutes after receiving it. He was not given any other medication prior to contrast medium containing diatrizoate meglumine 521 mg/ml and diatrizoate sodium 79 mg/ml with a total Iodine content of 292 mg/ml.

A Japanese case series (337,647 cases) reports that the overall risk of any adverse reaction was 12.66% with ionic

ICM and 3.13% with nonionic ICM. The author concluded that non-ionic agents were some 6-10 times safer than ionic agents.[4] Another study showed that 6000 patients who received ionic ICM, the incidence of mild adverse drug reactions was 2.5%; moderate reactions, 1.2%; and severe reactions, 0.4%.[5] However, in 7170 patients who received nonionic ICM, the incidences were only 0.58% for mild reactions, 0.11% for moderate reactions, and 0% for severe reactions.[6] Dillman et al performed a retrospective review of 11,306 children (age < 19 y) who received intravenous administration of low-osmolality nonionic ICM over 6.5 years period (January 1999 to June 2006) at their institution.[7] It was found that 0.18% of the children had acute allergic-like reactions to the contrast agent. Out of these children, 80% of the reactions were categorized as mild, 5% as moderate, and 15% as severe.

The use of steroids and antihistamines to prevent contrast material-induced anaphylaxis has been recommended by several authors but has not gained wide acceptance. [8,9,10] Data supporting the usefulness of premedication in patients with a history of allergic anaphylaxis are lacking and physicians who are dealing with these patients should therefore not rely on the efficacy of premedication. Antihistamines have a much slower onset of action than epinephrine, they exert minimal effect on blood pressure, and they should not be administered alone as treatment but should be considered adjunctive to epinephrine. Corticosteroids have no immediate effect on anaphylaxis, however, administer them early to try to prevent a potential late phase reaction (biphasic anaphylaxis). And with all these medication, early cardiopulmonary resuscitation is the key to patient's survival.

## Conclusion

This case illustrates how the administration of an intravenous contrast agent can have serious consequences in a susceptible patient. The absence of an anaphylactic reaction when test dose is given does not

preclude a life-threatening reaction from its subsequent administration in the same patient. And so, all precautions should be implemented and measures to treat serious reactions should be readily available.

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# Dysgerminoma With Pregnancy Presented With Hemorrhagic Shock

Shital Kapadia\*, Sumant Shah\*\*, Prerak Modi\*, Sandip Sonara\*\*\*, Nidhi Patel\*\*\*

## Abstract

Dysgerminoma (Germ cell tumor) is a rare tumor, rarer to be present with pregnancy, and it should be very rare for the operating surgeon to miss such a big mass. Germ cell tumors are basically found in pre-adolescent and adolescent girls. Here we present a case of dysgerminoma with pregnancy and highlight the need for appropriate management to prevent further complication when a rare pathology is encountered during pregnancy.

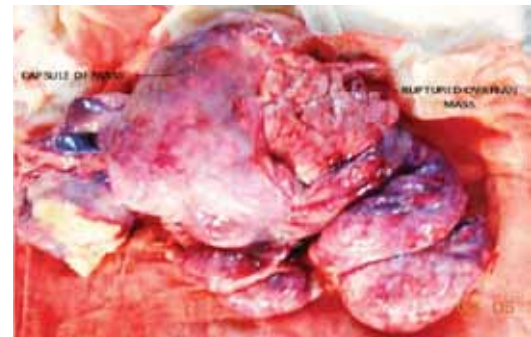
## Introduction

Ovarian dysgerminomas accounts for 2% of all ovarian malignancies and 28% of malignant germ cell tumors (GCT).<sup>1</sup> Malignant GCTs occur primarily in girls and young women.<sup>2</sup> They usually present as chronic abdominal pain and palpable abdominal lump with consequent distension. It may also present as an acute abdomen because of torsion of its pedicle.<sup>3</sup> A treatment consisting of surgical resection and platinum-based chemotherapy usually cures the majority of malignant GCTs.<sup>4</sup>

## Case History

A 30 years old second para female patient was referred from a private hospital where LSCS was performed for breech presentation 7 hours back. Patient was referred for bleeding from stitch (suture) line. On examination patient was pulse less and blood pressure was not recordable, respiration was regular. Patient had severe pallor. She was catheterised and urine output was 200cc. Abdominal drain was left by operating gynecologist showed 2.5 litres of frank blood in the bag. Per abdomen examination showed a 10 x 10cm mass palpable above and by side of the uterus. Per speculum and per vaginal examination showed no active bleeding. Patient was immediately administered 1 litre of Hemacel, one litre of ringer lactate and 500ml of Dextran 40. Blood transfusion through another intravenous line was started after cross match. Emergency ultrasound was performed. On ultrasound, uterus was bulky and there was a 13.9 x 10 cms sized homogenous echo textured lesion noted superior and lateral to the uterus with possibility of organised clot or broad ligament hematoma. Decision for

emergency laprotomy was taken. On laprotomy gross hemoperitoneum was present and 15 x 20 cms sized ruptured, fleshy, necrosed right ovarian mass was present which was source of active bleeding. The mass was removed and sent for histopathology examination. Patient was stabilized hemodynamically after 4 units of blood transfusion, 4 units of fresh frozen plasma and 6 units of platelets rich concentrate. But subsequently she developed anurea so she had to be shifted to kidney institute for hemodialysis. Histopathology of the specimen showed malignant germ cell tumor-dysgerminoma of the ovary with omental infiltration. Patient was advised chemotherapy after improvement in renal function.



## Discussion:

Dysgerminoma is the most common ovarian malignant germ cell tumor and can be found either in a pure or mixed form with other germinal elements.<sup>5</sup> The following histological subtypes of GCTs are reported: dysgerminoma, yolk sac tumor, embryonal carcinoma, polyembryoma, choriocarcinoma, immature teratoma, and mixed GCTs.<sup>4,6,7</sup> Patient usually present with chronic abdominal pain and palpable abdominal tumour.<sup>7</sup> It may present as acute abdomen because of torsion (tumor twisting around its pedicle) or intraabdominal hemorrhage (from congestion and rupture of superficial tumor vessels).<sup>3</sup> Some may present with vaginal bleeding, amenorrhea or precocious puberty.<sup>7</sup> Our patient had a rare presentation of dysgerminoma with pregnancy which was missed at the time of LSCS, done for breech presentation. Excessive bleeding resulted in hemorrhagic shock and subsequently renal failure. Generally surgery is the first step of management followed by adjuvant chemotherapy (depending on histopathology).<sup>7</sup> Appropriate surgical treatment for patients in whom fertility needs to be preserved consists of laparotomy with unilateral salpingo-oophorectomy and resection of all visible lesions, as done in this case.<sup>5,9</sup>

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The current standard adjuvant chemotherapy consists of 3 cycles of Bleomycin, Etoposide, Cisplatin (BEP protocol).<sup>6,9</sup> Four courses of BEP are recommended in case of bulky residual tumor after surgery.<sup>2,9</sup> Dysgerminoma is also very sensitive to radiation while other germ cell tumors are not.<sup>7</sup> Chemotherapy and/or radiation can be used to treat patients with metastatic disease.<sup>5</sup> For patients with early stage disease, cure rates approach 100%, while for those with advanced-stage disease it is least 75%.<sup>9</sup> Bilateral tumors occur in up to 10-15% of cases.<sup>4,5,7</sup> Recurrence in contralateral ovary can develop over the next 2 years.

## Conclusion

If patients with ovarian mass can be diagnosed earlier during the antenatal period or during the cesarean section than subsequent morbidities can be prevented. It is advisable to look for the possible causes in all patients with breech presentation.

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# Uterus didelphys, unilateral distal vaginal agenesis and ipsilateral renal agenesis : An unusual variant of OVIRA syndrome

Surbhi Gupta \*, Hafsa M. Vohra \*\*, Nilesh A. Shah \*\*\*

## Abstract :

We report a case of uterus didelphys with left hematometra, left hematosalpinx, left renal agenesis and arrested growth of ipsilateral vagina. It is believed that this case is a variant of the OVIRA syndrome : OVIRA syndrome is a rare unique clinical syndrome consisting of a double uterus, obstruction of the vagina (unilateral, partial or complete) and ipsilateral renal agenesis. The present case had symptoms of dysmenorrhea and lower abdominal pain because of obstruction as a result of arrested growth of vagina on the left side. The symptoms were relieved by excising the obstructed uterus, cervix and the fallopian tube.

**Key Words :** Uterus didelphys, ipsilateral renal agenesis, unilateral vaginal agenesis

## Introduction :

Abnormalities in the formation or fusion of the mullerian ducts can result in a variety of anomalies of the uterus and vagina : single, multiple, combined or separate. The close developmental relationships of the mullerian and wolffian ducts explain the frequency with which anomalies of the female genital system and urinary tract are associated. Disorders of lateral fusion of the two mullerian ducts can be symmetric unobstructed as with double vagina, or asymmetric obstructed as with unilateral vaginal obstruction. Obstructions associated with disorders of lateral fusion are particularly noteworthy in that they are observed clinically only as unilateral obstructions that almost invariably are associated with absence of the ipsilateral kidney. 40% of the female patients with congenital absence of kidney are found to have associated genital anomalies.<sup>(1)</sup>

## Case Report :

A 16 years old unmarried girl presented with complaints of occasional dull aching lower abdominal pain and dysmenorrhea since 1 year. She had attained menarche at the age of 14 years. Her menstrual cycles were regular lasting for 4-5 days every month with moderate blood loss

but excruciatingly painful.

On general and systemic examination, there were no significant findings. Her secondary sexual characters were well developed. Per abdomen examination revealed a palpable mass of about 4x4 cm size, firm in consistency in the left iliac fossa. Per speculum and per vaginum examination was not done. On per rectal examination, nothing conclusive could be assessed.

Hematological and biochemical tests were unremarkable. Ultrasonography reported uterus didelphys with two well developed uteri and cervix. Left side of uterus showed changes of hematometra possibly due to arrested development of left side of vagina. It suggested a remote possibility of bicornuate uterus or rudimentary left uterine horn with their non-communication with patent right side vagina. A complex fluid collection in left adnexa was observed which appeared to be a hydrosalpinx or chocolate cyst. The left kidney was absent with compensatory hypertrophy of the right kidney.

A CT scan suggested bicornuate didelphys uterus with hematometra and hematocolpos on left side associated with left hydrohematosalpinx and absent left kidney. The findings favoured mullerian duct anomaly which is shown as a schematic presentation in Figure -1.

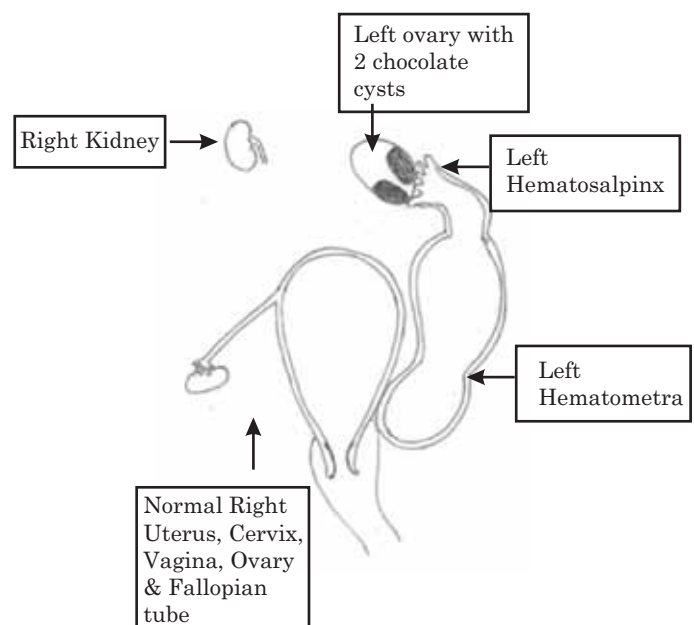


Figure 1: Schematic presentation of the case

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Intravenous pyelography (IVP) showed a non-functioning left kidney (absent left kidney-USG correlation) and normally excreting right kidney showing compensatory hypertrophy.

Exploratory laparotomy followed by removal of left non-communicating uterus and cervix and left salpingectomy was done. In the left ovary, two chocolate cysts were present and were removed. The residual normal left ovarian tissue was preserved. On intra-operative evaluation, a normal right uterus, right fallopian tube and right ovary could be seen. No rudimentary hemivagina, transverse or vertical septum was identified. The specimen was sent for histopathological examination and the architecture of endometrium, myometrium and cervix was confirmed (Figure-2).

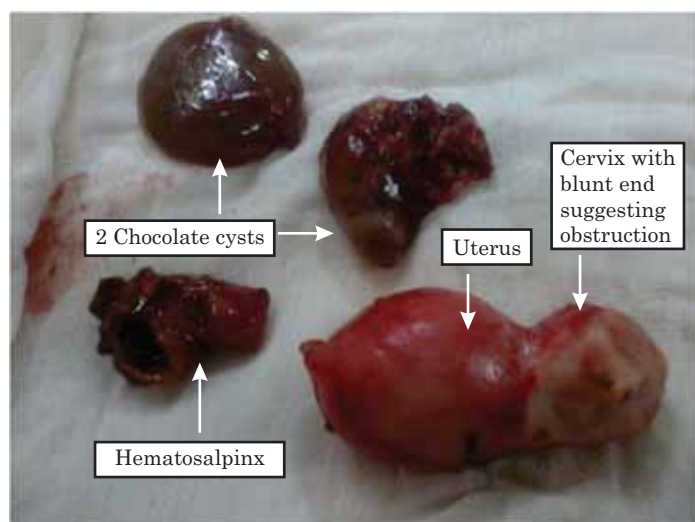


Figure 2 : Excised specimen-uterus with cervix with blunt end, hematosalpinx and 2 chocolate cysts

Section from specimen of tube showed architecture of fallopian tube with changes of chronic salpingoophoritis. The patient withstood the operation well and was discharged on 10<sup>th</sup> post-operative day. On follow up after 1 month, the patient was healthy with no complaints.

### Discussion :

The prevalence of mullerian duct anomalies in the general population ranges from 0.16-10% in various studies with one study employing a medline search showing a prevalence of 0.5%.<sup>(2)</sup> Mullerian duct anomalies have been classified in many systems over the years. As per the American Fertility Society classification of uterovaginal anomalies, this reported case would befit a variant of Class III A(iii), i.e. the lateral fusion defects-asymmetric obstructed disorder of the uterus or vagina-

with unilateral vaginal obstruction associated with double uterus – usually associated with ipsilateral renal agenesis. The patient had a class III Mullerian duct anomaly that demonstrated some features similar to OVIRA syndrome which presents with didelphys uterus, ipsilateral renal agenesis and a hemivagina. The cause of OVIRA can be related to damage of caudal portion of Wolffian duct. It is believed that the insult occurs from embryogenic arrest at 8 weeks of gestation that simultaneously affects mullerian and metanephric ducts.<sup>(3)</sup> Partial failure of fusion can result in single vagina with a single or duplicate cervix and complete or partial duplication of the uterus.<sup>(1)</sup> While a complete or a partial vaginal septum is associated with OVIRA in 75% cases, there was no vaginal septum in our case. It is a variant of OVIRA anomaly as there is no ipsilateral vaginal tissue. Clinically the spectrum of class III usually presents due to symptoms of obstruction. This presentation occurs at menarche when the hemiuterus becomes symptomatic and the patient experiences dysmenorrhea.<sup>(4)</sup> In our case, the right sided non obstructive uterus led to normal menstruation whereas the obstruction on the left side led to left hematometra with retrograde menstrual flow which led to hematosalpinx and was the cause of dysmenorrhea and lower abdominal pain.

Thus, because menses in patients with OVIRA syndrome are rarely irregular, the possibility of this syndrome as a diagnosis can easily be overlooked but a careful pelvic examination and the necessary radiological investigations will help to make the correct diagnosis. Hence this rare entity should also be considered while dealing with a case of mullerian anomaly.

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# Jarcho Levin Syndrome

Swati Upadhyay\*, Charul Purani\*\*, K.M.Mehariya\*\*\*

## Abstract :

Jarcho Levin Syndrome is a clinicoradiological entity characterized by short neck, short trunk, normal sized limbs and multiple vertebral and rib defects on skeletal survey. Severely affected individuals may have life threatening pulmonary complications due to deformities of thorax. This syndrome was first described by Saul Jarcho and Paul Levin in 1938. It has two clinically similar variants such as a Spondylo-thoracic dysostosis and spondylo-costal dysostosis. We report a case of 2.5 months old female child who had typical findings consistent with Jarcho Levin Syndrome, presented at Civil Hospital, Ahmedabad.

## Keywords :

Jarcho Levin Syndrome, spondylo-thoracic dysostosis, spondylo-costal dysostosis.

## Case History

A 2.5 months old female child presented with history of fever, cough and breathlessness since 15 days. She was the second child born out of non-con-sanguineous marriage. There was no history of significant antenatal complications. On general examination, patient had preauricular skin tag on right side, port wine stain on right arm and a supernumerary nipple on left side. (Figure -1).



**Fig.1: Child with Jarcho Levin Syndrome (spondylo-costal dysostosis) showing short neck, short trunk, preauricular skin tag and supranumerary nipple.**

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Abdomen was protuberant. On systemic examination, patient had tachypnoea, absent air entry on right side and prominent heart sounds on right side. The child weighed 4.1 kg, her length was 60 cm with an US: LS ratio of 1.36. On investigation, patient's hemogram was normal. Chest x-ray revealed homogeneous soft tissue opacity in right lung field with shifting of trachea, heart and mediastinum towards ipsilateral side, that was suggestive of loss of lung volume on right side. It also showed fusion of 1<sup>st</sup> & 2<sup>nd</sup> ribs, with crowding on right side. (Figure -2).



**Fig. 2: Chest X-ray showing fusion of 1<sup>st</sup> and 2<sup>nd</sup> ribs with crowding on right side, homogeneous soft tissue opacity in right lung field with shift of trachea, heart and mediastinum towards ipsilateral side.**

2D-echo showed dextrocardia and absent right pulmonary artery. Pulmonary angiography revealed thoracic vertebral anomalies (fusion of 1<sup>st</sup> and 2<sup>nd</sup> ribs on right side, fusion of posterior elements of D<sub>1</sub>, D<sub>2</sub>, D<sub>3</sub>, with spina bifida of C<sub>6</sub> and C<sub>7</sub> vertebrae, aplasia of right pulmonary artery and right lung field and single pulmonary artery. Ultrasonography of kidney, ureter, urinary bladder and brain were normal. The child was treated with IV antibacterial for 14 days and administered pneumococcal, H Influenzae type b and meningococcal vaccines on discharge to protect the normal lung from common pathogens, as the patient had right lung aplasia. Parents were counselled regarding the disease, its progression, associated complications and prognosis. Presently the baby is ten months old in perfect healthy condition.



## Discussion:

Jarcho Levin syndrome is a rare, genetic, axial skeleton growth disorder characterized by multiple vertebral and rib anomalies<sup>1</sup>. It was first described by Saul Jarcho and Paul Levin in 1938 at John Hopkins University<sup>2</sup>. The condition is characterized by reduced number of vertebral segments and the existing vertebrae exhibit a variety of anomalies including hemivertebrae, butterfly vertebrae and fused vertebrae associated with aplasia, hypoplasia and fusion of the ribs.<sup>3</sup> Our patient had fusion of 1<sup>st</sup> and 2<sup>nd</sup> ribs on right side, thoracic vertebral anomalies, neural tube defect and right pulmonary artery aplasia. Considering these findings the patient was diagnosed as second variant Jarcho Levin Syndrome i.e. spondylo costal dysostosis. Patients with Jarcho Levin Syndrome typically appear to have short trunk and neck with arms appearing relatively long with and protuberant abdomen. This patient had similar phenotypic observations. Hernias, Congenital heart defects, anomalies of the anal opening, urinary tract, external genitalia and lower limb are associated anomalies that may be present in this syndrome.<sup>4</sup> The genes involved in Jarcho Levin Syndrome are DLL3, MESP2, LFN4 and GDF6 mapping on chromosomes 19q13, 15q26.1, 7p22 and 8q22.1 respectively. The syndrome has two clinically similar variants such as spondylo thoracic dysostosis and spondylo costal dysostosis. Though both conditions are clinically similar they have different survival rates, associated anomalies and inheritance pattern. Spondylo thoracic dysostosis (STD) is inherited as an autosomal recessive disorder and has a grimmer prognosis than its counterpart. Spondylo costal dysostosis (SCD) is inherited as either autosomal recessive or autosomal dominant pattern. The survival rate is thrice than STD. In contrast to STD, the subtype SCD features intrinsic rib anomalies in addition to vertebral anomalies, which includes defects such as bifurcation, broadening and fusion that are not directly related to the vertebral anomalies. In STD, extensive posterior rib fusion occurs due to segmentation defects and extreme shortening of the thoracic vertebral column.<sup>5</sup> Prenatal diagnosis of Jarcho Levin Syndrome can be done by three dimensional ultrasound as early as 18 weeks of gestation.<sup>6</sup> Management aims at aggressive neonatal care, prevention and appropriate treatment of respiratory infections. Spinal surgery to improve the thoracic volume and decrease the pulmonary restriction has also been carried out. A correct diagnosis made in previously affected child can help in genetic counselling of couple at risk and establishing the diagnosis prenatally in subsequent pregnancy.

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## N.S.S. Annual Camps : Year 2011-12

With the help of District Health Authority, Ahmedabad total 7 one day camps were organized at different talukas of Ahmedabad by National Service Scheme (N.S.S) unit of B.J.Medical College, Ahmedabad during months of February and March 2012.



The camps were organized during non academic hours on Sunday / Saturday. The campaign was inaugurated at Rampura P.H.C. - Viramgam taluka by Dr.Parmar – R.C.H.O., Ahmedabad. Rampura, Kumarkhan, Modasar, Bagodara, Bhimnath, Saragnpur and Chaloda were selected for medical camps. Block Health Office (BHO) and concerned Primary Health Centre (PHC) selected the place for camps and made necessary arrangements including drugs.



Civil Hospital, Ahmedabad made arrangement for E.C.G. machine and other equipments for the camp. Transportation was arranged by Dean office of the college with the help of C.D.H.O., Ahmedabad.



This was a great opportunity for student volunteers to visit villages and participate in rural health awareness and health check up activity along with faculty members of civil hospital, Ahmedabad.



Experts from Medicine, Surgery, Gynec, Paediatrics, Psychiatry, Skin, ENT, Ophthalmology, Orthopedics participated in these rural camps. Blood bank of civil hospital arranged blood donation camp. Government Dental College extended support for dental OPD.

Around 1485 patients took benefits from these camps. Needy patients were referred to civil hospital.

Under the guidance of resident doctors of Community Medicine department, health awareness campaign was also arranged during camps. At each camp villagers were informed about various cancers, hazards of smoking and disease prevention activity through exhibition, skits and counseling.

A special screening programme for Diabetes was also arranged during each camp.

“It is different experience in camps where we see how our doctors examine and treat patients with minimum diagnostic support. The happiness in the eyes of villagers is unforgettable. It is my pleasure to participate and serve 'Real India' during my free time” says student volunteer.

Dr. Bharat Shah (Dean – B.J. Medical College), Dr. Geeta Kedia (Professor & Head, Community Medicine Department), Dr. Nilam Patel (C.D.H.O, Ahmedabad) and Dr. Shilpa Yadav (A.D.H.O., Ahmedabad) took active interest to make this camps possible.

Dr. Kriti, Dr. Meghavi, Dr. Pritesh and Dr. Krunal – resident doctors from community medicine department provided necessary guidance to volunteers during camps.

Dr. Dinesh Rathod (Programme Office – N.S.S. unit) coordinated medical camp activity.

## INSTRUCTION TO CONTRIBUTORS

### About the Journal

**BJ Kines**, a publication of B J Medical College & Civil Hospital Ahmedabad. The journal is presently biannually published by the college and available widely. The entire journal issue are available online free of cost from the web site of BJ Medical College. The journal does not charge for submission, processing or publication of manuscripts and even for colour reproduction of photographs.

### Scope of Journal

The journal will cover technical and clinical studies related to health, ethical and social issues relating to various aspects of medical science. Articles with clinical interest and implications will be given preference. The journal hopes to fulfil its mandate as the important journal of the state.

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Manuscripts accepted for publication are copy edited for grammar, punctuation, print style, and format. Page proofs are sent to the corresponding author. The corresponding author is expected to return the corrected proofs.

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Authorship credit should be based only on substantial contributions to each of the three components mentioned below :

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- Drafting the article or revising it critically for important intellectual content; and
- Final approval of the version to be published.

Participation solely in the acquisition of funding or the collection of data does not justify authorship. General supervision of the research group is not sufficient for authorship. The order of naming the contributors should be based on the relative contribution of the contributor towards the study and writing the manuscript. Once submitted the order cannot be changed without written consent of all the contributors.

### Conflicts of interest

All authors of must disclose any and all conflicts of interest they may have with publication of the manuscript or an institution or product that is mentioned in the manuscript and/or is important to the outcome of the study presented. Authors should also disclose conflict of interest with products that compete with those mentioned in their manuscript.

### Submission of Manuscript

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## **Types of Manuscript**

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These include randomized controlled trials, intervention studies, studies of screening and diagnostic test, outcome studies, cost effectiveness analyses, case-control series, and surveys with high response rate. The text of original articles amounting to up to 3000 words (excluding Abstract, references and Tables) should be divided into sections with the headings Abstract, Key-words, Introduction, Material and Methods, Results, Discussion, References, Tables and Figure legends. The Methods section should state whether the study has been approved by the institution ethics committee; and have details regarding study design, protocol, end points etc. Avoid duplication of information presented in tables / figures in text.

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It is expected that these articles would be written by individuals who have done substantial work on the subject or are considered experts in the field. The prescribed word count is up to 3000 words excluding tables, references and abstract. The manuscript may have about 90 references. The manuscript should have an unstructured Abstract (250 words) representing an accurate summary of the article.

### **Case reports :**

New, interesting and rare cases can be reported. They should be unique, describing a great diagnostic or therapeutic challenge and providing a learning point for the readers. Cases with clinical significance or implications will be given priority. These communications could be of up to 1000 words (excluding Abstract and references) and should have the following headings: Abstract (unstructured), Key-words, Introduction, Case report, Discussion, Reference, Tables and Legends in that order.

### **Letter to the Editor :**

These should be short and decisive observations. They should preferably be related to articles previously published in the Journal or views expressed in the journal. They should not be preliminary observations that need a later paper for validation. The letter could have up to 500 words and 5 references. It could be generally authored by not more than four authors.

### **References :**

References should be numbered consecutively in the order in which they are first mentioned in the text (not in alphabetic order). Identify references in text, tables, and legends by Arabic numerals in superscript with square bracket after the punctuation marks. Further guidelines can be download from <http://www.icmje.org> or [http://www.nlm.nih.gov/bsd/uniform\\_requirements.html](http://www.nlm.nih.gov/bsd/uniform_requirements.html)

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## Quiz!!

21 year old female known case of Seizure Disorder. She is not on any drugs. Her electrolytes are normal. ECG is suggestive of?



- a) Wolff-Parkinson-White (WPW) pre-excitation
- b) Long QT syndrome
- c) Hypokalemia
- d) Tricyclic overdose
- e) Hypothyroidism

**Answer on Page 18**



## Kaleidoscope of Events



**Dean Sir Hoisting The National Flag**



**Dean Sir Addressing The Audience During Independence Day Celebration**



**Gujarat University Youth Festival 2012 prize winners Ayushi Choksi, Preet Hathi, Mansi Satasia and Team.**



**Sarathak Joshi singing patriotic songs**

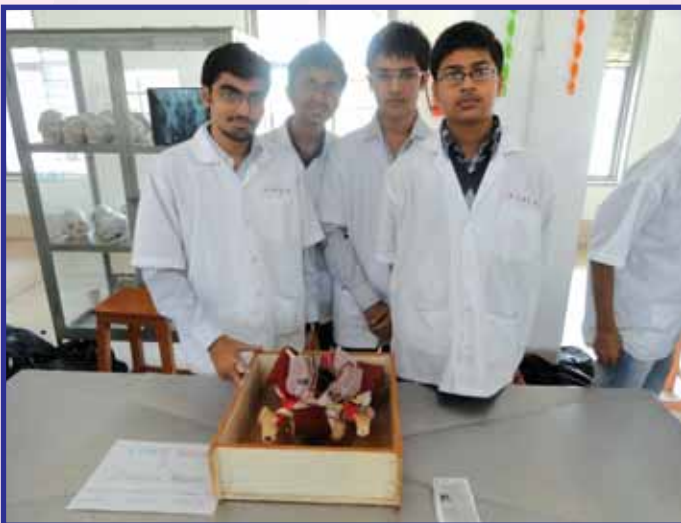
## Kaleidoscope of Events



Students with Prashman college wall magazine



Inauguration of chart and model competition by anatomy department



Prize Winning Model at the Competition



Dean sir at chart presentation



CME on ear organized by anatomy department



**Superbugs\* are visible manifestations of our prolonged failure to use antibiotics rationally**

## **Superbugs**

**Accumulation of resistance to multiple antibiotics**

**Self-medication and poor compliance**

**Inappropriate use of antibiotics leads to selection and multiplication of resistant strains**

**Weak surveillance and regulatory systems**

**Continuous natural evolution of resistance in microbes**

Known but neglected;  
need immediate action

Known but inevitable

\*Methicillin-resistant *Staphylococcus aureus*, MDR- and XDR-mycobacteria, ESBL producing gram-negative bacteria and NDM-1 producing enterobacteriaceae bacteria are a few examples of superbugs because they fail to respond to a large number of commonly used antibiotics.

**USE ANTIBIOTICS RATIONALLY**



*women's Hospital*

**Delivering | Life**

**Centre for excellence**

class-10,000 set-up  
IVF (test tube baby) ICSI  
freezing of embryos  
sperm and ovum donation  
surrogacy  
endoscopy  
fetal medicine  
menopause  
obstetrics  
gynecology  
health checkup



Dr. Pravin Patel



Dr. Manish Banker

**Orthodontia**

Braces for good smiles  
Lingual | Low-friction | Pre-adjusted

**Affiliations**

IVF: Melbourne IVF, Australia  
Endoscopy: CICE, France

**World class facilities**

deluxe suite & rooms  
postoperative ward  
three operation theatres  
conference room | auditorium