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Global Health and the 10/90 gap

Marco Luchetti

Global Health can be defined as "an area for study, research, and practice that places a priority on improving health and achieving health equity for all people worldwide". ¹ Article 25 of the 1948 Universal Declaration of Human Rights declares that, "everyone has the right to a standard of living adequate for the health of himself and of his family". ² Unfortunately, the health disparity between high-income and low-income countries, as well as between individuals within a country, often makes this impossible, leaving many people living in unhealthy settings without sufficient access to care.

The field of global health is concerned with the health of populations worldwide, focusing on issues that typically have global, political, and economic significance. These health issues usually transcend national boundaries and are best solved through international collaboration. ³ Global health initiatives aim to improve the health and wellbeing of impoverished, vulnerable, and underserved people worldwide. ¹ These initiatives include poverty reduction strategies, disease prevention measures (for HIV/AIDS, malaria, and tuberculosis, for instance), efforts to improve nutrition and food security, policy to raise environmental standards and living conditions, and the promotion of gender equality.

In 2001, the Commission of the World Health Organization (WHO) recommended to fund global health with 0.1% of GDP. 4

The average expenditure per capita for health in low-income countries is estimated at \$ 20 per year while that of Western countries is estimated at \$ 947. The target to be reached to help out the most disadvantaged countries is \$ 44-60 per capita, which would ensure the populations of the poorest countries with the access to essential health services. Directing the 0.1% of the GDP of developed Western countries to the aids for global health would mean closing the gap to reach the target base of \$ 44-60 that would allow the saving of 8 million lives a year. 4

Despite the good intentions, there is still a marked disparity between the current spending levels and the commitments made by developed countries in a context in which, among other things, the percentage of aid for global health has been in decline for almost all donor countries. Activists claim that only 10 per cent of global health research is devoted to conditions that account for 90 per cent of the global disease burden – the so-called '10/90 Gap'. ⁵ They argue that virtually all diseases prevalent in low income countries are 'neglected' and that the pharmaceutical industry has invested almost nothing in research and development for these diseases.

In fact, the WHO acknowledges that there are only three diseases that are genuinely 'neglected': African trypanosomiasis, leishmaniosis and Chagas disease. ⁶

A large proportion of illnesses in low-income countries are entirely avoidable or treatable with existing medicines or interventions. Most of the disease burden in low-income countries finds its roots in the consequences of poverty, such as poor nutrition, indoor air pollution and lack of access to proper sanitation and health education. The WHO estimates that diseases associated with poverty account for 45 per cent of the disease burden in the poorest countries. ⁷ However, nearly all of these deaths are either treatable with existing medicines or preventable in the first place.

If treatments exist for the majority of poor countries' health problems, why then do mortality rates remain so high? Any discussion of this question must address the problem of access to essential medicines, which remains an intractable political and economic problem. According to the WHO, an estimated 30 per cent of the world population lacks regular access to existing drugs, with this figure rising to over 50 per cent in the poorest parts of Africa and Asia. ⁸ And even if drugs are available, weak drug regulation may mean that they are substandard or counterfeit.

Within these populations, it is the poorest socioeconomic groups that disproportionately suffer from a lack of access to existing medicines. ⁹ The implications of this failure of public health policy on global mortality are profound – according to one study, over 10 million children die unnecessarily each year, almost all in low-income or poor areas of middle income countries, mostly from a short list of preventable diseases such as diarrhoea, measles, malaria and causes related to malnutrition. ¹⁰

Many governments fail their populations in this respect by imposing punitive tariffs and taxes on medicines, and by

skewing their spending priorities in favour of defence over health. The governments of poor countries hinder the creation of wealth, imposing obstacles in the way of owning and transferring property, imposing unnecessary regulatory barriers on entrepreneurs and businesses, and restricting trade through extortionate tariffs. It is these and other political failures that have left poor populations without the necessary resources to access the medicines that could so easily transform their quality of life.

In conclusion, it appears more and more urgent and necessary to decide where to direct our efforts and investment in research, without prejudice, analyzing all the possible strategies for tackling global health issues, including those standing beyond the current economic paradigm based on the market.

Competing Interests

None declared

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A Rapid Need Assessment Survey of Anaesthesia and Surgical Services in District Public Hospitals in Cross River State, Nigeria

Queeneth N. Kalu, Atim I. Eshiet, Essien I. Ukpabio, Anietimfon U. Etiuma and Emmanuel Monjok.

Abstract

Background: The district hospitals in Cross River State of Nigeria serve majority of the population residing in the rural areas and little is known about their capacity to provide anaesthetic and surgical care. The present study is an initial needs assessment of anaesthesia and surgical services in public hospitals in State stepping stone for strengthening anaesthesia and surgical as Methods: A standardized Lifebox Hospital Initial Needs assessment questionnaire and another structured questionnaire were used to assess the anaesthesia and surgical services in 16 general hospitals (district hospitals). The survey did not involve face-to-face interviews. Results: There was no practicing physician-anaesthetist in the 16 district hospitals in Cross River State. There were only 13 trained nurse-anaesthetists distributed unevenly in the 16 surveyed district hospitals. One visiting consultant anaesthetist and five visiting consultant surgeons from the University Teaching Hospital worked at the general hospital in the capital city leaving all others in rural locations without any specialist provision. Apart from the nurse-anaesthetists in the general hospital in Calabar and at Dr Lawrence Henshaw Memorial Hospital, also in Calabar both in the capital city, the others working in the rural hospitals have had no refresher or In-service training in the past 2 years. Many of the district hospitals lack basic anaesthetic equipment. An average of 3-53 surgeries are performed at the district hospitals with intravenous Ketamine as the commonest technique of anaesthesia. Conclusion: The district hospitals' anaesthetic and surgical capacity is grossly inadequate in relation to the population being served. It is recommended that shorter training programs for physicians at the primary and secondary levels of care and regular revision courses for nurse anaesthetists in Nigeria will strengthen the district surgical and anaesthetic capacity and services with urban-rural shift in manpower especially with government incentive for rural health workforce.

INTRODUCTION

Surgery and anaesthesia have traditionally been viewed as expensive, resource-intensive and requiring highly specialized training.1 This misconception has led to surgery and anaesthesia taking a back seat to public health, maternal and child health, and infectious diseases in global health.² Surgery has also been termed the "neglected stepchild of global health.3These concepts have changed rapidly since it has been found that surgical diseases contribute about 11% to Disability Adjusted Life Years⁴ and therefore would benefit from preventive and public health strategies necessary to achieve the Millennium Development Goals. The realization of the huge public health burden of surgical diseases in low and medium income countries (LMICs), and the fact that surgical services and treatment could be made cost effective, led World Health Organization (WHO) to launch the Global Initiative for Emergency and Essential Surgical Care (GIEESC) in 2005.5

The GIEESC is a global forum whose goal is to promote collaboration among diverse groups of stakeholders to strengthen the delivery of surgical services at the primary referral level in LMICs. Improvements in surgical services at the primary referral level in LMICs will equally require the provision of safe and effective anaesthesia. The provision of safe and effective anaesthesia will need adequately trained human resources and essential health technologies. The surgical and anaesthesia service capacity have generally been very low in sub-Saharan Africa (SSA) as evidenced through surveys conducted in

Ethiopia,6 Gambia,7,8 Ghana,9,10 Liberia,8,11 Malawi,12 Nigeria,13 Sierra Leone, 8,14 Rwanda, 15,16 Tanzania, 8,17 and Uganda. 18 The survey from Nigeria was conducted among rural private hospitals and was administered to attending members in a conference of the Association of Rural Surgical Practitioners of Nigeria.¹³ This was done using the Personnel, Procedures, Equipment and Supplies (PIPES) survey tool developed by the non-governmental organization Surgeons Overseas (SOS). 13 This is a tool developed to assess surgical capacity through the workforce, infrastructure, skill, equipment, and supplies of health facilities in LMICs.¹³ The other indicated surveys done in SSA used a comprehensive survey tool designed by the Harvard Humanitarian Initiative. This tool was adapted from the WHO Tool for Situational Analysis to Assess Emergency and Essential Surgical Care as part of an international initiative to assess surgical and anaesthesia capacity in LMICs.

The present survey used a rapid assessment tool known as the Lifebox Hospital Initial Needs Assessment questionnaire with another structured questionnaire to assess anaesthesia services in public hospitals in the Cross River State (CRS) of Nigeria. Lifebox (www.lifebox.org) is a non-profit organization saving lives by improving the safety and quality of surgical care in low-resource countries. Since 2001, Lifebox has trained more than 2000 anaesthesia providers, and more than 4200 pulse oximeters have been supplied to more than 70 low-resource countries thereby closing the operating room pulse oximetry gap in about 15 countries. This organization is supported by

the World Federation of Societies of Anaesthesiologists (WFSA), Association of Anaesthetists of Great Britain and Ireland, Harvard School of Public Health and the Brigham and Women's Hospital in Boston, United States of America. ¹⁹ This survey was primarily aimed at the secondary health care facilities which are owned and managed by the CRS Ministry of Health (MOH). This survey audit will identify the anaesthesia providers in CRS, their level of training and retraining as well as equipment available for providing safe anaesthesia and monitoring patients in the peri-operative period. The data will also identify baseline information and gaps in anaesthesia and surgical capacity as a first step for the CRS MOH initiative to improve surgical and anaesthesia services. This information is a stepping-stone for national and international assistance since CRS is a relatively poor state in the Nigerian Federation.

Country and State overview

Nigeria is the most populous African country, located in the West African sub-region with a population of more than 160 million people.20 It is a Federal Republic with 36 states and a Federal Capital Territory. It is politically sub-divided into six geo-political zones: North-Central, North-Eastern, North-Western, South-Eastern, South-South and South-Western. There are 774 Local Government Areas (LGAs) where more than 60% of the population reside. The health care system is divided into three levels: primary, secondary and tertiary. There are public and private health facilities operating at all levels. The primary healthcare facilities (health centres) are managed by the Local Government, the secondary healthcare facilities (general hospitals) are managed by the State Government, while the tertiary facilities (University Teaching Hospitals and Federal Medical Centres) are managed by the Federal Government. Health indicators for Nigeria are among the worst in the world despite the fact that Nigeria is the sixth largest exporter of crude oil. The United Nations Human Development Index ranked Nigeria 156 out of 187 countries.²¹ In particular, Nigeria is one of the five countries contributing more than 50% to the global maternal mortality ratio²² and one of the countries with the highest physician's and nurse's emigration to developed countries.²³ Physicians and nurses who remain in Nigeria predominantly practice in urban cities leaving the LGAs, most of them rural with severe shortages in health manpower.

CRS, with approximately 3.2 million population and 20156 square kilometres, is located in the South-South geo-political zone. ²⁴ The state has boundaries with the Republic of Cameroon in the East, Benue State in the North, Ebonyi State in the North West and Akwa-Ibom State in the South. ²⁴ It is divided into 18 LGAs with 18 general hospitals and 613 primary health centres. There is only one tertiary health facility, the University of Calabar Teaching Hospital (UCTH) located in Calabar, the capital city, which provides specialist care to the entire population. Being a tourism state, the importance of safe anaesthesia as a component of safe surgery cannot be overemphasized.

Physician-anaesthetist, nurse-anaesthetist and surgery training programs in Nigeria

Physicians are trained to be specialist anaesthetists or surgeons in a four-year training program leading to the Fellowship in Anaesthesia (FMCA) or Surgery (FMCS) of the National Postgraduate Medical College of Nigeria (NPMCN) or the Fellowship in Anaesthesia or Surgery (FWACS) of the West African College of Surgeons (WACS). This is after a six-year medical education program in the university leading to the Bachelor of Medicine and Bachelor of Surgery degree, one-year rotatory internship, and one-year of compulsory National Youth Service. Most Fellows, after completion (average time of completion is 7-8 years), work in University Teaching Hospitals and Federal Medical Centres, all located in urban cities. Another training program for doctors designed for primary and secondary healthcare is the Diploma in Anaesthesia (D.A) of the Universities or WACS, which is a 12-month training program. There is no short training program in Surgery.

Nurses are trained as nurse-anaesthetists after 18 months of training in a post-basic nursing program. The basic nursing training program is three-years of training in general nursing, after completing six years of secondary school education. There are now few university degree programs leading to the Bachelor's degree in Nursing Science (BSN) from the universities. All these nursing training programs lead to certification by the Nursing and Midwifery Council of Nigeria.

Rural-urban practice

Physician-anaesthetists (Fellows and Diplomates), nurseanaesthetists and consultant surgeons are all concentrated in urban hospitals leaving the rural areas and urban slums with a critical shortage of anaesthetic and surgical workforce. Therefore the majority of the surgical and anaesthetic procedures in rural areas in Nigeria are carried out by government-employed medical officers with almost all anaesthesia being provided by nurse-anaesthetists. In some very remote districts, Community Health Extension Workers (CHEWs) and Community Health Aids with little or no formal training in providing surgical care, are the only health workers available to provide some form of surgical care. The Association of Rural Surgical Practitioners of Nigeria (ARSPON) has been making some effort to address this workforce gap in rural areas by providing short on-the job training for medical officers to enable them to provide safe and affordable surgery to the rural population.¹³ The concept of surgical task-shifting to "nonphysician clinicians" to address this rural-urban surgical workforce disparity, as is being officially done in other LMICs of SSA²⁵ is not acceptable in Nigeria.

METHODOLOGY

A standardized questionnaire, the Lifebox Hospital Initial Needs Assessment Survey (Appendix 1) and another structured questionnaire (Appendix 2) was distributed to all 18 general

hospitals (secondary health facilities) in CRS. All the general hospitals, which are the first referral hospitals in the districts, perform surgery. The site visit was conducted in April/May 2014. Permission to conduct the site visit was given by the CRS Honorable Commissioner for Health. The hospital surveys did not involve face-to-face interviews with the medical superintendents, hospital matrons or anaesthesia providers. The questionnaires were to be completed by the anaesthesia providers and medical superintendents in each of the hospitals visited. Each completed questionnaire was to be sent to the office of the Honorable Commissioner for Health at the MOH headquarters in Calabar, the capital city. The results are presented in frequency tables and charts.

RESULTS

A total of 16 well-completed questionnaires were received from 18 general hospitals/secondary healthcare facilities visited (88.9 % response rate). Averages of 3 - 53 surgeries are performed monthly in each of the hospitals (Table 1). The common procedures performed include: herniorrhaphy, appendicectomy, caesarean section, myomectomy, prostatectomy and exploratory laparotomy (Table 1). There are no practicing physician anaesthesiologists or surgeons employed by the State MOH except for one visiting consultant anaesthesiologist and five visiting consultant surgeons from the University of Calabar Teaching Hospital (UCTH), at the General Hospital, Calabar, which is located in the capital city of the State (Table 1). There are 13 nurse-anaesthetists distributed unevenly in the 16 hospitals (Table 1). There are no clinical officers cadres in the Nigerian healthcare system. Apart from the nurse-anaesthetists at the General Hospital in Calabar and Dr Lawrence Henshaw Memorial Hospital, also in Calabar, the other nurseanaesthetists have had no refresher course or in-service training in the past two years. In the 16 general hospitals, the commonest anaesthetic technique used is Total Intravenous Anaesthesia (TIVA) with Ketamine (Table 1).

Basic anaesthetic equipment such as anaesthetic machines, oxygen cylinders, suction machines, and pulse oximeters were lacking in most of the hospitals visited (Box 1).

The WHO Surgical Safety Checklist Information was administered to the hospitals management team at the Districts Hospital (Box 2). This shows that all the surgical teams had never used the WHO checklist, never received training, and the checklist was not available in the operating rooms, although all surgical personnel would like to receive training on the WHO checklist and pulse oximetry

DISCUSSION

This survey aimed to provide a quick assessment of anaesthesia and surgical services in public hospitals in CRS of Nigeria. The data shows gross and significant shortages in anaesthesia and surgical providers in all 16 general hospitals. There were no consultant anaesthetists, diplomate anaesthetists or consultant

surgeons employed in the CRS MOH. There were only 13 nurse-anaesthetists working in the 16 general hospitals, and one visiting consultant anaesthetist and five visiting consultant surgeons at the General Hospital, Calabar, the capital city. In six of the hospitals, there were no nurse-anaesthetists providing care for the surgical procedures being conducted. As it has been reported from the many surveys in SSA.^{6, 18, 26} most of the procedures in all the hospitals are being done by generalist medical doctors and general nurses, many without any postgraduate training in surgery and anaesthesia.

The gross inadequacy of the anaesthetic workforce in this survey represents what is found in many of the 778 LGAs (Districts) in the Federal Republic of Nigeria. This is because many of the LGAs are rural and studies have indicated the general difficulties of most health workers seeking jobs in rural hospitals. The lack of specialist anaesthetists in peripheral hospitals in most of Nigerian Districts therefore requires a redirection of the training programs for doctors in Nigeria with greater emphasis on the shorter training program design for primary and secondary healthcare levels. Inaddition, annual refresher courses should be made mandatory for nurse-anaesthetists especially for those practicing in rural areas.

A recent review of the met and unmet needs of surgical disease in rural SSA, where district and rural hospitals are the main providers of care, shows a very huge burden.²⁷ An important finding is the discrepancy between surgical care needs and provision.²⁷ Since the majority of the population in SSA reside in rural areas, there is the need to strengthen the surgical services at this level. This is the first of the four recommendations of the Bellagio Essential Surgery Group.²⁸ Many of the surveys using the WHO Situational Analysis Tool have described the lack of capacity in many district hospitals to meet the local surgical and anaesthesia needs. 6-18 One study, using pulse oximeter availability as a measure of operating room resources, showed that between 58.4% and 78.4% of operating rooms in West Africa, East Africa and Central SSA do not have pulse oximeters.²⁹ This finding was also clearly shown by our own rapid survey and assessment. Three important factors have been responsible for these findings. These are lack of resources, lack of manpower and the need for training.²⁷ The need for training to improve the quality of the surgical and anaesthesia providers at the district hospitals is the third recommendation of the Bellagio Essential Surgery Group.²⁸

Training programs and improvement of the facilities at the District Hospital has been shown to increase the number of operations performed.²⁷Also, the presence of a visiting consultant anaesthetist in the District Hospital has been shown to increase the scope of anaesthesia services during the visiting period.³⁰ The visit left more knowledgeable local staff in the care of their patients especially in peri-operative care.³⁰ The need for developing countries in SSA, particularly in Nigeria, to concentrate more on shorter training programs in surgery and

anaesthesia at their current level of development has been advocated. ^{31,32} This has been shown by Sani et al where a 12-month training program for General Practitioners in district hospitals in Niger significantly reduced the number of referrals to the regional and specialist hospital. ³³ In many other SSA countries, where gross shortages of medical manpower exist, surgical task shifting has been championed and research has shown that these are cost effective interventions. ^{25, 34} This is, however, not acceptable in Nigeria which is Africa's most populous country with very poor health indicators.

There are some limitations to this study. Firstly, it was a snapshot of anaesthesia and surgical services which did not highlight in detail the eight key areas of surgical and anaesthesia care, as in other surveys. These key areas include: access and availability of hospital services, human resources, physical infrastructures (including availability of water and electricity), surgical and anaesthetic procedures, surgical and anaesthesia outcome, essential equipment availability, NGO and international organizations providing care, and access to essential pharmaceuticals. Secondly, this assessment did not include the only public tertiary hospital in the State and private hospitals. Lastly, this was an initial assessment in preparation for a more detailed survey based on the WHO guidelines when research funds are received.

CONCLUSION AND RECOMMENDATIONS

There has been a paradigm shift in global public health and the concept of primary healthcare which has resulted in increased awareness of the importance and contributions of surgical disease to the overall burden of disease especially in LMICs. This rapid survey of anaesthesia services in CRS, one of the 36 states in Nigeria, will serve as a window to inform other Nigerian State governments of the need to increase surgical and anaesthesia capacity and funding in their development agenda. It is therefore recommended that visiting consultant's services to all the general hospitals in an organized and planned fashion should be highly encouraged. All the anaesthesia caregivers should attend refresher courses at least once every two years. These courses can be arranged locally, or sponsorship provided for attendance of relevant courses by Anaesthesia Trainers within and outside the State. Basic anaesthesia equipment and guidelines as recommended by the Nigerian Society of Anaesthetists (Box 3) must be available and followed to enhance patient safety. It is also recommended that the government should give incentives to medical and nursing staff working in rural areas so that there will be a reversal of the rural- urban shift. The Lifebox global oximetry project is interested in making high quality, low-cost pulse oximeters available in every operating room. Therefore, every secondary care facility in the country should take advantage of this laudable program.

Table 1: Summary of audit of anaesthesia services in the 16 district hospitals in Cross River State, April/May2014

Hospital	Number of anaesthetists		Type of anaesthesia	Scope of surgery	Average	Refresher	
	Nurse- anaesthetist Physician- anaesthetist		administered		number of surgeries	course within last two years	
General Hospital, Obubra	0	0	Ketamine	Herniorrhaphy, Laparotomy, C/S, Appendicectomy,	10	No	
General Hospital, Ogoja	4	0	GA(ETT), Spinal, Local, Ketamine	Herniorrhaphy, Laparotomy, C/S, Appendicectomy, Myomectomy & Others	15	No	
General Hospital, Sankwala	1	0	Local	Others	11	No	
General Hospital, Okpoma	0	0	Local, Ketamine	Herniorrhaphy, Appendicectomy & Others	3	No	
General Hospital, Calabar	1	1 (Visiting)	GA (ETT), Face Mask, Spinal, Epidural, CSE, Local, GA(Ketamine)	Herniorrhaphy, Laparotomy, C/S, Appendicectomy, Myomectomy & Others	53	Yes	
General Hospital, Ugep	1	0		Herniorrhaphy, Laparotomy, C/S, Appendicectomy, Myomectomy & Others	18	No	
Cottage Hospital, Oban	0	0	Ketamine	Herniorrhaphy, C/S, Appendicectomy, Myomectomy	4	No	
Cottage Hospital, Akpet	0	0	Local, Ketamine	Herniorrhaphy, Laparotomy, C/S, Appendicectomy.	10	No	
Lutheran Hospital, Yahe	1	0	Local, Ketamine	cal, Ketamine Herniorrhaphy, Laparotomy, C/S, Appendicectomy, Myomectomy & Others			

Eja Memorial Joint Hospital, Itigidi	1	0	Local, Ketamine	Herniorrhaphy, Laparotomy, C/S, Appendicectomy, Myomectomy	15	No
St. Joseph Hospital, Akpabuyo	1	0	Local, Spinal, Ketamine	Herniorrhaphy, C/S, Appendicectomy, Myomectomy	5	No
Dr. Lawrence Henshaw Memorial Hospital, Calabar South	1	0	GA (ETT), Face Mask, Local, Ketamine	Herniorrhaphy, Laparotomy, Appendicectomy, Myomectomy & Others	10	Yes
General Hospital, Ukem, Odukpani	0	0	Ketamine	Herniorrhaphy, Appendicectomy	3	No
Ranch Medical Center, Obudu	1	0	Ketamine	Herniorrhaphy, Laparotomy, C/S, Appendicectomy, Myomectomy	5	No
General Hospital, Akamkpa	1	0	Local, Ketamine	Herniorrhaphy, Laparotomy, C/S, Appendicectomy, Myomectomy	20	No
Government House Clinic, Calabar	0	0	Local, Ketamine	Herniorrhaphy, Laparotomy, Appendicectomy	3	No
TOTAL	13	1 (Visiting)			190	

GA: General Anaesthesia. ETT: Endotracheal Intubation. C/S: Caesarean Section. CSE: Combined Spinal and Epidural anaesthesia.

Box 1. Summary of Equipment in the 16 General Hospitals, Cross River State, Nigeria: April-May 2014

- · 10% of the hospitals had pulse oximeter
- · 20% of the hospitals had oxygen cylinders
- · 20% of the hospitals had suction machines
- \cdot 30% of the hospitals had anaesthetic machines
- \cdot 80% of the hospitals had recovery beds
- · 100% of the hospitals perform surgery

Box 2. World Health Organization (WHO) Surgical Safety Checklist Information

- · How often do the surgical teams at your hospital use the WHO Surgical Safety Checklist? NEVER
- · Has your hospital received training in the WHO Surgical Safety Checklist? NO
- · Is the WHO Surgical Safety Checklist available in your operating rooms? NO
- · Would you like to receive training in pulse oximetry and the WHO checklist? YES

Box 3. Nigerian Society of Anaesthetists Standard Guidelines for the Practice of Anaesthesia

Anaesthetic Personnel

- · Certified physician anaesthetist
- · Trained nurse-anaesthetist under supervision by physician-anaesthetists
- · Maximum number of nurses to physicians should be 4:1
- · Where there is no physician-anaesthetist, nurses should adhere strictly to the guidelines and conditions of their certification
- · The surgeon should provide coverage especially in the area of patient resuscitation and fitness for surgery and take full responsibility for any decisions made against the guidelines.

Anaesthetic equipment for each theatre

Standard Continuous Flow (Boyles) Anaesthetic Machine with:

- Closed breathing system
- Adult semi-closed breathing system
- Paediatric breathing system

Suction Machine

- Electric
- Manual

Suction Catheters – disposable, various sizes

Laryngoscope set with batteries and:-

- 2 standard and 1 long curved blades

- 2 standard and 1 long straight blades
- Neonatal laryngoscope with 2 straight blades.

Intubating Forceps (Magill)

- Adult
- Paediatric
- Neonatal

Self-inflating Resuscitation Bag

- Adult
- Paediatric
- Infant

Anaesthetic Face Masks : Size 0, 1, 2, 3, 4 Paediatric (Rendell-Baker) – Size 00,0,1

Naso-gastric tubes Head – Harness

Oropharyngeal Airways 00 – 5

Endotracheal tubes (cuffed, noon-cuffed 2.5. – 9.0. mm)

- Red rubber, latex reinforced, portex

Plastic laryngeal Mask Airway (Sizes 1-5)

Endobronchial tubes

Bougies

Fluid Warmer

Warming Mattress (for paediatrics)

Pressure Infusor

Syringe Infusion Pump with lines

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Competing Interests

None declared

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Version 1/April 2012

Appendix 1



Hospital Initial Needs Assessment Survey

INSTRUCTIONS: Please fill out the form as completely as possible. Required fields are highlighted in red when using Adobe Reader. If there are required fields which you are unable to complete, type any response but make a note in the Additional Comments section. Once the form has been completed, save the form using the Save As button and e-mail the form to info@lifebox.org.

Today's Date:
Your Contact Information
First Name: Last Name:
Job Title:
Email Address: Mobile Number:
Hospital Information
Hospital Name:
Mailing Address
Street Address:
City: Dept/State/Region:
Country: Mailing Code:
Type of Hospital: Governent Private Other
Hospital Human Resources
SURGERY: Please indicate the number of people performing surgery in your hospital.
Trained Surgeon General Physician Clinical Officer Other
ANESTHESIA: Please indicate the number of people providing anesthesia in your hospital.
Trained Anesthesiologist Clinical Officer Nurse Anesthetist Other
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Appendix 2

Questionnaire for the survey of public hospitals in Cross River State, Nigeria.

	IT OF ANAESTHETIC SERVICES IN CROSS RIVER STATE se tick appropriate Box(s) or insert number
(1)	NAME OF HOSPITAL:
(2)	NUMBER OF ANAESTHETISTS:
(3)	Please state the designation of Anaesthesia Provider(s): Dr(Fellow) Diploma in Anaesthesia Nurse Anaesthetist Others please specify: GENERAL PRACTITIONER
(4)	Please indicate the number of Permanent/Visiting Staff in boxes provided: Permanent Staff Visiting Vi
(5)	Please any Refresher Course Attended in last 2 years?:
(6)	Type of Anaesthesia administered: GA(ETT) Face Mask Spinal Epidural CSE GA(TIVA e.g. Ketamine)
(7)	No. of Surgeons: General O&G Orthopaedic Others
(8)	Average number of surgeries done monthly:
(9)	Total number of surgeries done in past 6/12:
(0	Scope of surgeries done: (a) C/S (b) Hernionhaphy (c) Appendicectomy (d) Laparotomy (e) Myomectomy (f) Others
	Do you manage patients with these conditions? Hypertension Asthma Sickle Cell ase Diabetes Mellitus Elderly Paediatrics Neonates
Summ	nary of available anaesthesia equipment Equipment needs
	<u> </u>

Prevalence and implications of genital tattoos: A site not forgotten

Thomas Neluis, Myrna L. Armstrong, Cathy Young, Alden E. Roberts, LaMicha Hogan and Katherine Rinard

Abstract

Purpose: To provide information on men who have tattooed one anatomical site, the genital region (pubic and/or glans penis), that is uncommonly noted.

Methods: Two methods were used. First, the limited cultural and medical literature was reviewed. Secondly, a subsample of 14 men were analyzed, taken from a primary study examining male genital piercings (N = 445), who responded affirmatively to one survey question about penile tattoos. Findings: The literature (n = 25) was limited. Cultural literature revealed a long, rich history of genital markings for esthetics, sexual enhancement, and tribal status, whereas the medical literature reflected limited observational type information, some actual case histories, and few studies. From the small subsample, qualitative and quantitative data were provided. Similarities to those who wore general body tattoos were validated such as being single, heterosexual, having some college/vocational education, monthly binge drinking, no skin complications, and a strong propensity for a Need for Uniqueness. Reportedly, they were major body art wearers and continue to enjoy them. First age occurrence of sexual intercourse was similar to the national average of 17 years. Challenged assumptions included (a) no consensus regarding being risk takers, (b) significant reported forced sexual activity, and no (c) physical, sexual or mental abuse.

Conclusions: From our experience, those with genital tattoos are seen primarily for a normal range of developmental and physiological urologic issues, not their decorative markings; these genital tattoos are an integral part of their cultural and personal expression and most likely will increase. Yet, the markings are only skin deep so clinicians should adopt a nonjudgmental approach and employ methods of proactive patient health education.

Keywords: Key Words: Penis, tattoos, males, genital tattoos, Need For Uniqueness

INTRODUCTION

Maintaining its longstanding presence as one of the oldest forms of art, body tattooing has increased exponentially within mainstream society, as well as in social acceptance. Generally worn to display individuality and creativity, these distinctive forms of indelible markings are present in every culture, whether on tribal men, or people of status. Procedurally when inserting the decorative markings, the approach in studio tattooing has not changed significantly as artists are still using "an electrically powered, vertically vibrating instrument to inject tattoo pigment 50 to 3,000 times per minute up to or into the dermis at a depth of $1/64^{\text{th}}$ to $1/16^{\text{th}}$ of an inch"¹

While no national registry provides prevalence, a 2012 Harris Poll cited one in five United States (U.S.) adults have at least one tattoo (21%), an increase of 16% and 14% from previous surveys taken in 2003 and 2008 respectively. Tattoo numbers were even higher in some variables including age between 30-39 years (38%), Hispanics (30%), females (23%), and those living in the Western part of the U.S. (26%). No questions were identified in the 2012 poll that queried tattooed body site locations. Other studies cite almost a 25% presence of tattoos. The amount of tattoo studios also echoes the growing body art phenomenon.

Given the societal blaze of tattooing, the medical literature on body art has also increased. Yet, most of the information still remains focused on small case reports⁶ about traditional locations (arms, legs, chest, back), their decision-making,

various risk-taking behaviors,⁸ and the small amount of complications.⁷ Those with various adverse skin reactions or major complications seem to have had tattoos with colored pigment.⁶

While body art can be found virtually everywhere on the human anatomy, several articles have surfaced concerning genital body piecing. 4-5, 9-11 Current studies validate the increasing rate of all types of tattooed 4,8 people, from a variety of occupations and social classes, with markings on visible and non-visible locations.7 This article reports on the limited medical literature found about men with genital tattoos (pubic and/or the glans penis). Also presented is a subsample data analysis of 14 men from a primary study examining male genital piercings, 11-12 who responded affirmatively to one survey question about penile tattoos. This synopsis and subsample data analysis are provided for clinicians to have further, recent evidence about men with genital tattoos for decision making during patient encounters in health care settings. The terminology of penile and genital tattoos will be used interchangeably in this article.

METHODS

Literature Synopsis

Historically, the cross-cultural literature is rich in visual genital tattoo descriptions. In South America, the Moche on the North Coast of Peru (A.D. 150-800) produced ceramics illustrating vivid sexual imagery and highly decorated male genitals.¹³ Phallus decorations with dots, concentric lines, and

other tattoo markings on the penile skin and mucosa during the Upper Paleolithic era in Europe 12,700 to 11,000 years ago have been reported. 14-15 Likewise, the Samoan Island culture, where the word "tattoo" is believed to have originated from "tatau,", has maintained ritualistic16 traditions for over two thousand years; they are initiated at the time of puberty for future leadership roles. These 10+ days of ceremonies include very painful, repeated tattooing of the scrotum (tafumiti) and the penis (tafito). Other nearby primitive Polynesian tribes have believed this tattooing as highly erotic, 16 whereas the indigenous Maori (New Zealand) trust that the pigment for these tattoos can trap cosmic energy.¹⁴ Circumcision and tattooing were thought to produce the same effect of magic protection and healing powers after scar healing.¹⁴ In the Japanese culture, an examination of Yakuza (racketeers or gangsters) also describes the genitalia as a site that is tattooed, 17 fulfilling their principles of tattoos always being covered.

Searching for information about genital tattoos was more challenging within the medical literature. A comprehensive longitudinal 40 year search of the national and international electronic medical literature (1973-2013) published in English and their associated reference lists was conducted with MEDLINE, EMBASE, CINAHL, SCOPUS, and OVID. Only 20 articles were located that mentioned genital tattoos. Articles were from international authors (n = 11) and the U.S. (n = 9); they all produced interesting reading. One reference cited women with genital tattoos.⁷

Genital tattoos in the early literature were labeled as criminal, or personality disorders tattoos;18 one recent article discussed them under the header of genital self-mutilation.¹⁶ Others described them as a valuable clue for forensic pathology identification. 19-²⁰ World War II articles cited descriptive stories of soldiers with penile tattoos, 21-22 with one reporting up to 10 sailors being seen.²³ Besides reporting on how the fate of Bulgaria was determined by three tattooed men (Churchill with an anchor on his left arm, Roosevelt with a family coat of arms tattoo, and Stalin with a death's head on his chest),²⁴ Kazandjieva²⁵ then provides vivid examples of auto-aggression markings that his countrymen self-inflicted after the Communist takeover. This included glans penis tattoos which are described as producing great pain. 15,25 One political candidate, while campaigning, is reported as suggesting punitive action for those HIV+ by "putting indelible, glow-in-the-dark tattoos on [their] genitals."26 Traumatic tattoos associated with gunpowder explosions and blast burns are also mentioned on the glans penis.27

Two studies also described inmates with genital tattoos and discussed how these markings demonstrated aggressive behavior within this type of environment. Here large, colorful tattoo designs and wording on the glans penis tattoos were described²⁸⁻²⁹ which seemed to satisfy the inmate's flaunt of personal pain endurance. Additionally, Cuban refugees (Marielitos) fleeing to the U.S. were reported as having genital tattoos; they also were

Four other reports described those with penile tattoos also routinely inserting foreign bodies^{12,30} and paraffinoma^{12,31,32}into the penis. In Pehlianov's study (also in Bulgaria) they included a control group of another 25 men with genital tattoos. Recently, a unique case of non-ischemic priapism for 3 months was reported³³ following prolonged bleeding from a manual penile tattoo procedure in Iran. The authors suggested the hand-held tattoo needle had penetrated too deeply producing an arteriovenous fistula and the subsequent persistent half-rigid priapism. The authors also noted that the 21 year old patient expressed no regret, depression, or other complications related to the genital tattoo.

Original Study

The initial study queried males with genital piercings using available internet survey software, ¹² as it was considered a hidden variable. Anonymity and access to people nationally and internationally were major advantages for using this nontraditional approach. The university institutional review board deemed the study status as Exempt. To obtain quantitative and qualitative data about those men with genital piercings, an 83 item web-based survey was used; overall results, and another subsample of this data, are published elsewhere. ¹¹⁻¹²

Subsample of those with Penile Tattoos

From the original 445 male genital pierced individuals that responded to the question regarding having tattoos on their penis, 14 replied affirmatively. This subsample had previously been determined not be an outlier of the larger group of genital pierced men. While a short general description (age span at the time of tattoo procurement, urethral "play," design types, motives, and tattooists) about the 14 member genital tattoo subsample was published in 2010, 12 further investigation leading to quantitative and qualitative (Figure 1) data is presented here.

Figure 1: Subsample Respondent Qualitative Quotes

- *Black tribal flames on the top of the shaft, done at [age] 38
- * For erotic reasons, self done with no complications, done at [age] 54
- *I got it because I wanted it. After it was finished I realized I needed it, done at [age] 30
- * I self tattoo'd my penis on the glans and around the corona ridge in order to make up for its' lack of size and to enhance its appearance. I used a sailmaker's needle and Indian ink and there were no complications, done at [age] 43.
- *one small cross pigment tattoo!
- *I'm a little more than average in size, but I still have issues with my genitals. The way they look and their size. Piercings and tattoos have helped me quite a lot.

*I sketched a rose one day, like[d] the design, decided to get it tattooed on my penis. The stem is green with some yellow highlights, the bud is red, all black outline. The tattoo was applied with a standard machine . . .healing was actually quicker and easier than any of my other tattoos.

*It's a little heart just next to 'captain hemingway' which I hand poked and used india ink for it when I was 17... thought our penis deserved a reminder of our affection . . .no complications experienced but since it was hand done with a [sterile] needle it's kind of blurry

This subsample had significantly more foreskin genital piercings (chi-square = 11.5) = 1; P = .001), whereas the most common genital piercing of the larger group of those without genital tattoos¹¹ had Prince Albert piercings (inserted through external urethra). No question inquired which came first, the genital piercing or genital tattoos.

Data Analysis

For this subsample analysis, (and original study¹¹⁻¹²), IBM SPSS 21was used to obtain frequencies and chi-square analysis. Cross tabulations for the subsample were obtained by comparing those with and without penile tattoos.

RESULTS

Demographics

Almost all of the subsample respondents with penile tattoos were reportedly Caucasian (92%) and their ages ranged from 18 to 67 years (average 42.3). Of those that replied, six lived in the U.S. and five cited various international locations. Over half had vocational or college education (64%) and significantly more were likely to be single (25%) or divorced (25%), (chisquare 12.6) = 5; P = .027). Data regarding religious faith was weak to non-existent (75%). Respondents self-reported a good state of health (92%) (chi-square = 8.7) = 3; P = .034), yet 50% cited no annual health check-ups.

Risk Behaviors

Within this subsample, there was no consensus about being a "risk taker". Recreational drugs were reportedly not used (91%), over half were non-smokers (55%), but monthly alcohol use with binge drinking (5+ or more drinks) was cited (78%). Their "motives for genital tattoos were for esthetics, sexual, and personal pleasure" a variety of penile tattoo designs were described (Figure 1), created either "by studio artists (n = 11) or self-inflicted (n = 3)". All of them described having other body art, such as piercings and other general body tattoos. Some reported an average of 4 piercings (81%) and a significant amount of general body tattoos (average 3.5) (chi-square = 11.1) = 5; P = .049), that still interest them (85%) (chi-square = 8.9) = 3) P = .031).

Sexual Activity

This subsample's average age of first intercourse was 17 years, with most citing women as their sexual partners (92%), most preferred penile/vaginal intercourse (79%), and only one respondent reported a sexually transmitted infection (gonorrhea). When asked about any forced sexual activity (rape), this subsample had a significant amount of those who answered affirmatively (23%) (chi-square = 7.7) = 1; P = .005). Virtually no sexual, physical, or mental abuse was reported.

Need for Uniqueness

A four-item scale called the Self-Attributed Need for Uniqueness (SANU)³⁴ was present in the survey to determine the respondent's self-view (Cronbach alpha = .86). Using a Likert scale, the subsample's moderate, strong and very strong perspectives were collectively summarized. These respondents with penile tattoos preferred to be different (79%), distinctive (86%), intended to do things to make themselves different than those around them (72%), and reported a Need For Uniqueness (93%) (Cronbach alpha = .77). To validate this finding, when all 5 responses of SANU were totaled,¹² the mean was 12.43 documenting a more positive perspective for intentionally wanting to be different, distinctive, and unique.

DISCUSSION

This article reviewed both the cross cultural and medical literature about those with genital tattoos, as well as included both a quantitative and qualitative subsample data analysis of a small group of men who specifically reported penile tattoos. Yet, with certainty this small sample size produced limitations and reporting/survey bias. Additionally, any generalizability with the findings of this subsample should be noted as the respondents could have self-selected their participation and used their personal judgment to interpret the survey questions in this non-experimental cross-sectional study using internet survey methodology.¹²

From this review and to our knowledge, few have studied groups of men with genital tattoos, a difficult group of subjects to find with this hidden variable. 12,31 Cultural descriptions documented a long, rich history 12, 14-17, 29,31 of genital markings for esthetics, sexual enhancement, and tribal status, whereas the medical literature reflected limited observational type information, and few actual case histories or scientific studies. Although there were no mental health evaluations 12 cited in this medical literature, more psychopathic, deviant behavior discussions were made about the individuals with genital tattoos. 16-18,26,32,35 In contrast, two authors 30,33 comment on the "normalcy" of their patients that presented with genital tattoos.

Genital tattoos may be more common than this very small subsample size suggested as great emphasis has been placed on male penile size in many cultures, for a long time.^{31,36} The augmentation of these genital markings and decorative designs

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seemed to have motivated their sexual health, self-enhancement⁹⁻¹⁰ and well-being.³¹ Thus, when further studies are considered for this population with a hidden variable, these findings should assist with further ideas of investigation.

Current society has a strong 25 year renaissance of procuring tattoos with at least one in five, and perhaps even four, individuals possessing a tattoo, on virtually every part of their body, without major complications. This small subsample of those who have genital tattoos validates some similarities to those who wear general body tattoos such as a single heterosexual orientation, possessing some college/vocational education, monthly binge drinking, 1,3-5,10 and a strong propensity for a Need for Uniqueness. 4-5,37 They were major body art wearers and continue to enjoy them, as others have also reported. 4-5,10-12

Yet other demographic assumptions were challenged for this subsample of men with genital tattoos. These international respondents tended to be older Caucasians and not as ethnically diverse; there also was not a consensus as to them being risk takers, as has been repeatedly reported by many other body art respondents. 1,3-5,11-12

Subsample respondents reported their average first occurance of sexual intercourse at age 17, similar to the national figures.³⁸ Significant experiences of rape were also reported in this subsample, as in women with genital piercings.^{4-5,9-10}The national rate for forced sexual activity is 10.5%³⁸ and those with genital tattoos reported over twice that amount (23%). No sexual abuse was reported in contrast to a recent German study³⁹ examining general body tattooing.

As with any type of invasive procedure, there can be complications with certain types of body art. When these complications occur, body art wearers typically first seek the internet and/or their studio artist for health advice before presenting to clinicians. 1,8,10-12 Yet, overall for the amount of general tattooing done, this type of body art produced limited documented complications and more potential concerns. 7-8,11, 30,33 More complications were reported when the tattoos contained colored pigments. 6

These tattoos are an integral part of their cultural and personal expression. ^{12,31,33} From our experience many of these male patients with genital tattoos are not seen primarily because of their decorative markings, ³¹ but during clinical evaluations for other issues presented with the normal range of urologic issues involving overall genitourinary and sexual function. Genital tattoos can be an ambivalent findings for many clinicians, but these indelible skin markings (tattoos) are only skin deep, ⁴⁰ and provide valuable cues such as a history of sexual trauma. ³⁹ Currently more genital tattoos are seen among our freedom-impaired patients, where the prevalence of general body tattoos among the inmates can be as high as 67%. ⁴¹

Anecdotally, when healthcare staff discover a patient with genital body art, this discovery can be met with judgmental attitudes and behaviors which could impact care. To adequately assess, evaluate and treat the individuals that have chosen to have genital tattooing, clinicians should strive to provide a thoughtful, nonjudgmental patient-centered approach, along with a generous application of health education, for their present, or even future body art.¹¹

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Competing Interests

None declared

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Association between plasma adiponectin and risk of myocardial infarction in Asian Indian patient with diabetes

Arun Narayan, Sanjay Kulkarni, Rahul Kothari, Telugu Seetharam Deepak and Punith Kempegowda

Abstract

Context: Recent epidemiological studies have established association of adiponectin with insulin resistance and cardiovascular risk factors. However, newer reports state an ethnic difference in this association.

Objectives: The present study was done to assess the association between plasma adiponectin levels and coronary event in Asian Indian patients with diabetes. The relation between plasma adiponectin and various cardiovascular risk factors in an acute coronary event was also studied.

Methodology: The prospective study was conducted at a tertiary care center in Bangalore, India. Three groups of 30 patients-Patients with diabetes with Myocardial Infarction (MI), Patients with diabetes without MI and controls (age and sex matched non-patients with diabetes)- were included in the study. The association between plasma adiponectin level and MI in patients with diabetes was studied in comparison to patients with diabetes without MI.

Statistical analysis used: Analysis of Variance, Spearman Correlation

Results: Patients with diabetes with MI had significantly lower plasma adiponectin when compared to patients with diabetes without MI which in turn was lower than in normal subjects (P<.001). Plasma adiponectin was significantly correlated with abdominal obesity (r=-.31), fasting glucose level (r=-.61), glycated haemoglobin (r=-.63) and triglycerides (r=-.54) (all P<.001). There was no significant correlation between plasma adiponectin levels and High Density Lipoprotein-Cholesterol in the present study.

Conclusions: The present study and the recent evidence suggest that cross-talk between inflammatory signalling pathways and insulin signalling pathways may result in insulin resistance and endothelial dysfunction that synergize to predispose to cardiovascular disorders.

Key Messages: Adiponectin is a potential target in future research for reducing morbidity and mortality associated with atherosclerotic disease.

Keywords: Adiponectin, Diabetes, Myocardial infarction, HDL cholesterol.

Introduction

Adiponectin, first reported in 1995 by Scherer et al, is a novel and important member of the adipokine family. It is a collagen-like protein that is exclusively synthesised in white adipose tissue and is the gene product of adipose most abundant gene transcript 1 (apM1).

Adiponectin has been postulated to play an important role in the modulation of glucose and lipid metabolism in insulinsensitive tissues in both humans and animals. Various studies have reported a protective effect of plasma adiponectin against type 2 Diabetes Mellitus (T2DM) ²⁻⁶. Adiponectin is also inversely associated with traditional cardiovascular risk factors, such as total and low-density lipoprotein cholesterol (LDL-C) and triglyceride levels, and is positively related to high-density lipoprotein cholesterol (HDL-C).⁷ Recent studies suggest that it may have anti-atherogenic and anti-inflammatory properties .⁸⁻¹⁰ A few researchers who studied the combined effects of these findings reported inverse correlation between plasma adiponectin and risk of coronary heart disease.¹¹⁻¹⁵

Recent epidemiological studies have shown that association of adiponectin with insulin resistance and cardiovascular risk factors vary with ethnicity. Mente et al studied the ethnic variations in adiponectin concentrations and insulin resistance and found that South Asians and aboriginal people display a greater increase in insulin resistance with decreasing levels of

adiponectin compared to Chinese and Europeans .¹⁶ However, a similar study involving Asian Indian teenagers showed that adiponectin did not correlate directly with measures of insulin sensitivity, overweight, and other cardio-metabolic variables .¹⁷ Similar studies in adults are not available.

The present study was done to assess the association between plasma adiponectin levels and coronary event in patients with diabetes. Also the relation between plasma adiponectin level and various cardiovascular risk factors were studied in patients with diabetes with and without acute coronary event.

Subjects and Methods:

The prospective study was conducted at a tertiary care centre in Bangalore, India from January 2008 to December 2009. The study was approved by the institution ethics committee. Three groups of patients, age and sex matched, were included in the study. The first group included 30consecutive T2DM patients admitted with a diagnosis of myocardial infarction (MI) at the study centre. While the second consisted of patients with T2DM without MI, the third group were patients without diabetes without any history of acute coronary event. MI was diagnosed per World Health Organization's criteria.18 Patients aged less than 18 years were not included in the study. Patients with diabetes with chronic kidney disease or receiving Thiazolidinediones were also excluded from the study as it would alter plasma adiponectin levels.

Fasting Blood Glucose (FBG), Post-Prandial Blood Glucose (PPBG), Glycated Hemoglobin (HbA_{1C}), Fasting Lipid Profile, Baseline Electrocardiogram and Plasma Adiponectin were done for all the study subjects. In addition, Coronary Angiogram was done for patients with diabetes with MI to confirm Coronary Artery Disease (CAD) and treadmill tests were done for patients with diabetes without MI to exclude underlying CAD.

FBG and PPBG, serum total cholesterol and serum triglycerides were estimated using enzymatic kit method (Vital Diagnostics, Mumbai, India); and serum HDL-C (Bayer Diagnostics, Baroda, India) using a semi-auto-analyser.

Plasma Adiponectin levels was estimated using Human Total Adiponectin/Acrp30 Quantikine ELISA Kit (R&D Systems Inc., India). This assay employs the quantitative sandwich enzyme immunoassay technique. A monoclonal antibody specific for the Adiponectin globular domain has been precoated onto a microplate. Standards and samples are pipetted into the wells and any Adiponectin present is bound by the immobilised antibody. After washing away any unbound substances, an enzyme-linked monoclonal antibody specific for the Adiponectin globular domain is added to the wells. Following a wash to remove any unbound antibody-enzyme reagent, a substrate solution is added to the wells and colour develops in proportion to the amount of Adiponectin bound in the initial step. The colour development is stopped and the intensity of the colour is measured.

Statistical Analysis

Statistical analyses were performed using Statistical Package for Social Sciences (SPSS) for Windows 16.0 (SPSS Inc., Chicago, USA). The results for each parameter (numbers and percentages) for discrete data and average (mean ±standard deviation) for continuous data are presented in tables and figures using Microsoft office 2007 software package.

Two-way Analysis of Variance (ANOVA) was performed for plasma adiponectin in patients with diabetes with MI, patients

with diabetes without MI and controls as the grouping factor. Two tailed 'P' values less than 0.05 were considered significant. Spearman correlation was performed to analyse the association between plasma adiponectin, BMI, FBG, PPBG, HbA_{1C}, serum triglycerides, HDL-C and LDL-C.

Results

The following results are expressed as mean± standard deviation. The mean age of the study subjects in the three groups-patients with diabetes with MI, patients with diabetes without MI and Controls- was 58.00±8.77 years, 57.17±9.34 years and 54.20±7.28 years respectively. The descriptive statistics of the various parameters under study is given in table 1.

Patients with diabetes with MI had significantly lower plasma adiponectin levels (6.11±1.82) when compared to patients with diabetes without MI (9.47±1.55) which in turn was lower than normal subjects (17.82±1.30) (P<.001) . Plasma adiponectin was significantly correlated with BMI (r=-.31), FBG (r=-.61), HbA_{1C} (r=-.63) and triglycerides (r=-.54) (all P<.001). We did not find any significant correlation between plasma adiponectin levels and HDL-C (Table 2).

Discussion

In the present study, we found decreased plasma adiponectin concentrations in the patients with diabetes which was further lower in patients with an acute coronary event indicating that it may be a predictor of macroangiopathy. Hotta et al found similar results in their study and proposed that accumulation of adiponectinin atherosclerotic vascular walls may accelerate its half-lifein plasma, resulting in the reduction of the plasma concentration of adiponectin in subjects with CAD .¹⁹ Ouchi et al studied the molecular basis of link between adiponectin and vascular disease and found that adiponectin modulates endothelial inflammatory response and that the measurement of plasma adiponectin levels may be helpful in assessment of CAD risk. ²⁰

Table 1: Descriptive statistics of various parameters under study

Variable	Controls Mean (±Std. Dev)	Patients with diabetes without MI Mean (±Std. Dev)	Patiente with diabetee with
Plasma Adiponectin	6.11(±1.82)	9.47(±1.55)	17.82(±1.30)
Fasting Blood Glucose	123.50(±17.85)	133.23(±16.14)	88.80(±6.27)
Post-Prandial Blood Glucose	190.53(±19.27)	209.33(±28.72)	125.30(±6.200
Glycated Haemoglobin	7.81(±0.92)	8.04(±1.24)	4.06(±0.62)
Total Cholesterol	205.77(±19.92)	214.43(±21.54)	138.07(±10.38)
Serum Triglycerides	148.80(±11.32)	160.53(±14.61)	127.23(±6.11)
Serum HDL	45.13(±8.57)	37.43(±9.73)	44.87(±7.78)
Serum LDL	129.30(±22.55)	137.27(±18.83)	120.03(±8.27)
Body Mass Index	27.82(±2.39)	27.08(±2.20)	25.40(±2.63)

Patients with diabetes with MI had significantly lower plasma adiponectin when compared to patients with diabetes without MI which in turn was lower than in normal subjects

Table 2: Spearman correlation between adiponectin and body mass index, blood lipids, HbA1C, fasting and post-prandial glucose levels

	Adiponectin	BMI	FBG	HBA1C	TG	HDL	LDL
Adiponectin	1.00	-0.31**	-0.61**	-0.63**	-0.54**	0.02	-0.16
BMI		1.00	0.37**	0.29**	0.25*	-0.14	0.10
FBG			1.00	0.62**	0.61**	-0.17	0.21*
HBA1C				1.00	0.83**	-0.45**	0.35**
TG					1.00	-0.53**	0.33**
HDL						1.00	-0.07
LDL							1.00

^{**} Correlation is significant at the 0.01 level (2-tailed); * Correlation is significant at the 0.05 level (2-tailed); (BMI- Body Mass Index, FBG- Fasting Blood Glucose, HDL- High Density Lipoprotein, LDL-Low Density Lipoprotein, HBA1C- Glycated Haemoglobin, TG- Serum Triglycerides); Plasma adiponectin was significantly correlated with BMI, FBG, HbA1C and triglycerides (all P<.001). The correlation between plasma adiponectin levels and HDL-C was not statistically significant.

Large scale prospective experimental research is needed to clarify these theories.

The relation between plasma adiponectin and the various known metabolic risk factors were on par with the world literature, except for HDL-C. Koenig et al reported an additive effect of HDL-C and adiponectin on CAD risk prediction. ²¹ In their joint analyses, the highest risk for T2DM as well as acute coronary events was observed in men with low adiponectin in combination with low HDL-C levels. In the present study, the mean HDL-C levels were lower in patients with diabetes with MI compared to patients with diabetes without MI. However, we did not find any significant correlation between plasma adiponectin levels and HDL-C in the present study. Similar findings were obtained by Schulze et al indicating that although plasma adiponectin has been established to be correlated with insulin resistance, CAD and metabolic disease, the interrelation between these is far more complex.

The molecular mechanisms by which adiponectin exerts its multiple functions and whether its actions are receptor mediated still remain a mystery. Is the primary activity of adiponectin antiatherosclerotic, or is it principally a modulator of lipid metabolism and regulator of insulin sensitivity, or is it all of the above? The answers to these and other intriguing questions will undoubtedly provide additional insight into the metabolic roles of this new adipocyte hormone.

Conclusion

The present study and the recent evidence suggest that cross-talk between inflammatory signalling pathways and insulin signalling pathways may result in insulin resistance and endothelial dysfunction that synergise to predispose to cardiovascular disorders. Large scale prospective studies are needed to examine the ability of increase in adiponectin levels and insulin sensitivity to improve primary end points including incidence of diabetes and outcomes of cardiovascular events.

Competing Interests

None declared

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Severe Presentation of Acute Upper Airway Obstruction – A Case Report

Adeel Majeed and Asquad Sultan

Abstract

Tongue swelling (glossitis) can be caused by many conditions. We present a case of severe tongue swelling leading to severe, acute upper airway obstruction and its anaesthetic management in the emergency setting.

Keywords: Airway Obstruction, Angio-oedema, Anaphylaxis, Tracheostomy,

Abbreviations: A&E - Accident and Emergency, IV - intravenous, IM - intramuscular, O2 - oxygen, GCS - Glasgow Coma Scale, ICU - Intensive Care Unit, ENT - Ears Nose and Throat, ALS - Advanced Life Support, CPR - Cardio-pulmonary Resuscitation, ETT - endotracheal tube

Case

A 74 year old female presented to the A&E department after waking with a swollen tongue. She called for an ambulance and the paramedic crew initially treated her for an allergic reaction with 200mg hydrocortisone IV, 20mg chlorphenamine IV and four doses of 0.5mg adrenaline 1 in 1,000 IM. She did not improve and was transferred to the local A&E.

In A&E, she was initially stable with no stridor or difficulty in breathing but noticeably swollen tongue. Saturations were 98% on 5L nasal O_2 . Blood pressure was stable at 135/75 mmHg. GCS was 15/15, but the patient was agitated from not being able to speak or retract the tongue. No further history was taken other than an allergy to shellfish, with no recent exposure. An obese habitus was noted.

A&E doctors called the anaesthetic on-call team, and on their request called the on-call ICU consultant and the on-call ENT consultant. Further adrenaline 100mcg IV, hydrocortisone 200mg IV and chlorphenamine 10mg IV were given. An attempt to look inside the mouth with a tongue depressor and torch was made by the anaesthetics/ICU team and it was quickly realised that the swelling continued into the mouth and larynx and was rapidly progressing. Given the lack of suitable equipment and the severity of the case, a decision was made to transfer to emergency theatres.

Once in emergency theatres, the on-call ENT consultant was scrubbed and ready. 100% oxygen via facemask and routine monitoring was instituted. At the request of the ENT consultant, a micro-tracheostomy was attempted with local anaesthesia but failed to pass into the trachea given the patient's habitus. The ENT surgeons attempted an awake tracheostomy, but this was difficult due to her being agitated and unable to lie still, and a calcified trachea.

The rapidly progressing swelling compromised oxygen delivery to the lungs and the saturations began to drop quickly. The patient became bradycardic and lost consciousness. At this point, it became easier to attempt the tracheostomy. The ALS protocol was followed and CPR started. An attempt was made for direct laryngoscopy – a grade 3b view was obtained and a size four microlaryngeal tube was passed successfully. 100% O₂, two doses of 1mg adrenaline IV, and 3mg atropine IV were given and the heart rate improved. Pulses were present and the defibrillator showed sinus rhythm; CPR was stopped and tracheostomy was continued. Due to abundant peri-tracheal fat, a number of tracheostomy tubes were tried before a secure tracheostomy was placed. However the microlaryngeal tube maintained airway patency.

An arterial line and larger cannula was secured. Propofol infusion and fentanyl IV were given to maintain anaesthesia and the patient was transferred to ICU.

On ICU, there was further difficulty in ventilation, with high airway pressures and saturations falling to the low 90s despite 100% O₂. It was thought the tracheostomy tube was abutting the carina or posterior tracheal walls. The ENT surgeons were called urgently and in the interim the patient was re-intubated with a size 7 ETT to maintain the airway. The ENT team changed the size 8 cuffed non-fenestrated tracheostomy tube for a size 7 Shiley with a proximal extension. Despite the change, ventilation remained difficult. An urgent chest X-ray was performed which showed a right-sided pneumothorax. A chest drain was inserted and a 'hiss' was noted on insertion, indicating a possible tension pneumothorax. Ventilation then improved.

Further history was taken from previous notes and discharge letters. It was noted that the patient had allergies to shellfish, penicillin, erythromycin, and diclofenac, but no history of

exposure. She had a background of hypertension, angina, chronic kidney disease and previous breast cancer. She was taking Lisinopril, Diltiazem, Nicorandil, Tamoxifen, Sertraline and Omeprazole. Her tongue swelling improved markedly over the next two days with antibiotic treatment and regular dexamethasone. She was awakened once appropriate and haemodynamically stable. A nasendoscopy was performed by the ENT team on day six and nothing remarkable was noted, with swelling having regressed totally. She was decannulated on day eight and transferred to the ward. Lisinopril was stopped.

Discussion

This case demonstrated a severe, acute presentation of tongue swelling (glossitis) leading to upper airway obstruction. Although a number of conditions may cause glossitis: infection, trauma, anaemia, liver disease, malnutrition. Acute glossitis is a hallmark of angio-oedema. This is a rare, but life threatening condition that requires prompt recognition and treatment.

Angio-oedema may result from anaphylactic, hereditary, acquired or idiopathic processes. Some 12-24% of anaphylaxis cases present with angio-oedema.^{1, 2} Hereditary and acquired cases usually result from a deficiency of C1 esterase inhibitor deficiency, which causes an accumulation of bradykinin, leading to soft tissue oedema. Such an increase in bradykinin may be caused by angiotensin converting enzyme inhibitors, leading to angio-oedema.³ If no cause can be found, it is termed idiopathic.

Management of angio-oedema requires rapid airway assessment and management; resuscitation; and treatment of the underlying cause. Anaphylaxis should respond to standard management as outlined by the AAGBI.⁴ Angio-oedema from other causes requires cessation of the suspected causative agent³, and in an emergency, nebulized adrenaline to reduce airway swelling. Infusion of plasma derived or recombinant C1 Esterase Inhibitor may also rapidly improve symptoms.¹

Although airway support is the cornerstone of anaesthetic management, an acute, rapidly progressing case such as this requires a multi-disciplinary approach. Anaesthetists will require help from operating department practitioners and nurses to manage the initial airway compromise. A compromised airway presents a significant hazard to any form of anaesthesia, especially if it results in cardio-respiratory depression, which will expedite hypoxemia and impair tissue oxygenation. An awake, spontaneously ventilating approach to secure the airway needs to be undertaken. ENT staff should be scrubbed and ready to perform an emergency surgical tracheostomy if complete airway obstruction occurs or airway access cannot be secured.

An awake fibre-optic intubation can be attempted but this requires an experienced anaesthetist, timely access to equipment, preparation and a co-operative patient. These are unlikely to be provided in the resuscitation room. Fibre-optic

manipulation causing bleeding or further swelling can lead to complete airway obstruction.

An inhalational induction to maintain spontaneous ventilation and then followed by direct laryngoscopy or fibre-optic intubation is another option and reduces the required cooperation of the patient. But this may cause haemodynamic instability in an already compromised patient and can lead to complete airway collapse.

An elective awake tracheostomy under local anaesthetic is the most likely route to ensure airway access without haemodynamic compromise. This will require a co-operative patient, senior help from trained operating department staff, and the ENT surgeons scrubbed and ready to perform a surgical tracheostomy if a percutaneous approach fails.

The Intensive Care Unit should be aware of the patient and ICU teams will be required to help with airway access as well as manage haemodynamic instability, secure arterial and central venous access. The patient will need further airway support and treatment in the Intensive Care Unit.

An acute upper airway obstruction therefore requires an emergent, yet controlled, approach to secure airway access and maintain oxygenation. Staff should have clear roles, ideally decided beforehand and practiced, with experience in the use of the equipment available. Senior ENT help should be available readily and again be versed in securing an emergency awake tracheostomy should other means fail or are not suitable.

As with any critical incident a debriefing should be undertaken to highlight points in the management of such patients that were handled well and those that were not, so that existing management plans can be improved and skills honed to improve management of future incidents.

Competing Interests

None declared

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Cutaneous Polyarteritis Nodosa: A case report

Harish J, Manjunath M N and Chaithanya C Nair

Abstract

Cutaneous polyarteritis nodosa is a rare vasculitis of childhood relating to small-to-medium-sized arteries. Its etiology is unknown. Clinical manifestations include tender subcutaneous nodules, livedo reticularis, cutaneous ulcers and necrosis. Although it is distinct from systemic polyarteritris nodosa in that it lacks significant internal organ involvement, extra-cutaneous manifestations may be evident. The diagnosis is by skin biopsy as there is no other specific serological tests and characteristic pathologic feature is a leukocytoclastic vasculitis in the small to medium-sized arterioles of the dermis with or without fibrinoid necrosis. Mild cases may resolve with nonsteroidal anti-inflammatory drugs. If more severe, treatment with systemic corticosteroids generally achieves adequate response. We report a 10 year old girl with cutaneous PAN, who presented to us with arthralgia and swelling of left knee joint and both ankle joints and fever , with multiple tender subcutaneous nodules on both upper and lower limbs.

Keywords: Cutaneous polyarteritis nodosa, subcutaneous nodules, vasculitis.

Abbreviations: cPAN- Cutaneous polyarteritis nodosa, PSM- pansystolic murmur. CRP- c reactive protein, ASLO- anti streptococcal lysin O

Polyarteritis nodosa (PAN) is a rare vasculitis in childhood. Since first described by Kussmaul and Maier in 1866 ¹, there have been approximately 140 pediatric case reports in the literature. Traditionally, children were classified as having one of three forms: infantile, cutaneous, and systemic. Infantile PAN is now recognized as a severe form of Kawasaki disease. Criteria for a diagnosis of systemic PAN in childhood have been proposed but not validated².

Cutaneous PAN (cPAN) is recognized as a separate entity but there are no diagnostic criteria for cPAN². cPAN is characterized by disease affecting the skin with no major organ system involvement. The cutaneous symptoms are similar to systemic PAN and mild fever, muscle, joint, and peripheral nervous system involvement may also occur. Fever, rash, and musculoskeletal symptoms are common in children and cPAN needs to be differentiated from other diagnostic entities. Definitive diagnosis is by histopathologic evidence of necrotizing inflammation of the medium and small-sized arteries. There is a paucity of knowledge of the spectrum of clinical presentation and management of children with cPAN. Here we describe a case of cPAN and summarize the clinical manifestations, laboratory data and treatment regimens of our patient.

Case report

This 10 year old female adolescent presented with pain in both the elbow joints followed by pain in the left knee joint and both the ankle joints in a course of 8 days and fever for the past 2 days. On admission her vitals were stable, both the elbow joints were tender and the knee and ankle joints were swollen and tender. She had multiple subcutaneous nodules over extensor aspect of both her forearms, both her tibial shins and few on her thighs. Systemic examination showed presence of a soft PSM of

grade 2 intensity over left sternal edge. Blood investigations showed leucocytosis, elevated CRP, elevated ASLO titres and 2Decho revealed a mild tricuspid regurgitation. An initial diagnosis of acute rheumatic fever was made and child was started on penicillin and Aspirin. But child continued to have excruciating arthralgia and hence a rheumatologist opinion was taken. Child was advised a skin biopsy from the nodular lesions which showed small and medium vessel vasculitis suggestive of cutaneous polyarteritis nodosa.

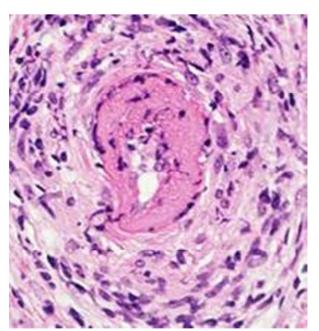


Fig 1 showing segmental fibrinoid necrosis with inflammatory infiltrates of small artery.

Hence she was stopped with aspirin therapy, given pulse therapy with methyl prednisolone and continued with penicillin therapy. Her arthralgia subsided within a day of pulse therapy and the subcutaneous nodules gradually disappeared. On

discharge child was put on oral steroid therapy and penicillin prophylaxis and advised regular follow up.

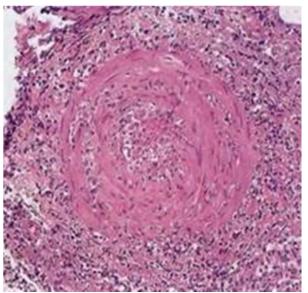


Fig 2 showing leukocystoclastic vasculitis with fragmentation of neutrophils in and around blood vessels.

Discussion

cPAN is not common in the pediatric population with approximately 140 cases reported in the literature. Disease is limited to skin, joints, and muscles in the majority with a minority having nerve involvement. Constitutional symptoms are common. Most children have a chronic and relapsing benign course.

The precise etiology of cPAN remains to be unknown. However, an immune mediated mechanism has been postulated. Several infectious and noninfectious conditions have been associated both to initiation and relapse of the disease^{3,4,5}. Among them, streptococcal infection has been commonly implicated^{6,7}. Although some evidence of streptococcal infection as an initiating factor for cPAN is present, caution must be exercised when interpreting elevations in the serologic markers of streptococcal infection in the absence of an appropriate clinical presentation.

Cutaneous and systemic PAN share the same histopathologic features of necrotizing arteritis of small and medium sized vessels. Kussmaul and Meier described the first case of systemic PAN in 1866 ¹. Early reports ^{8,9} confirm that cPAN is a separate entity to systemic PAN. We have limited our definition of cPAN to disease affecting the skin, muscle, joints, and peripheral nervous system, with corresponding biopsy confirmation. Any evidence of visceral involvement, either clinically (central nervous system, pulmonary, cardiac, gastrointestinal, or renal), radiographically (abnormal angiography), or by histology (visceral biopsy) were classified as systemic PAN. Nakamura et al ¹⁰ proposed further restriction of the definition of cutaneous PAN in that any extracutaneous involvement such as peripheral neuropathy and myalgias must

be limited to the same area as skin lesions. Systemic PAN and cPAN appear to be fairly distinct entities on a clinical continuum. There are only 5 reported cases of cPAN evolving into systemic PAN 11,12 .

On review of treatment regimens reported in the literature, most children respond to corticosteroids. Penicillin should be considered in children with increased ASO titres ^{13,14}. Recent case series report success with low-dose methotrexate, cyclophosphamide, intravenous immunoglobulin, and biologic therapies ^{15,16}.

In summary, cPAN can be challenging to diagnose and manage. A diagnosis of cPAN should be considered in a child with fever, tender subcutaneous nodules, livido reticularis, and arthralgias/arthritis. Most children respond to corticosteroids and have a benign course, but some children may be corticosteroid dependent or corticosteroid resistant, necessitating other immunosuppressive agents including DMARDs and biologic therapy. Multicentre pediatric vasculitis disease registries are necessary to inform development and standardization of best clinical practice for childhood cPAN.

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None declared

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Case report: DiGeorge syndrome presenting with hypoparathyrodism and Learning Difficulties in adulthood

Nawras Al-taie, Sandra Scheuter-Mlaker, Michael Schlesinger and Heidemarie Abrahamian

Abstract

We report a 40 year old female with mild dysmorphic facial features, learning difficulties, epilepsy and chronic dermatitis, presenting with symptomatic hypocalcaemia. The laboratory investigations confirmed the diagnosis of hypoparathyroidism. The hint to DiGeorge syndrome was the hypoparathyroidism in association with learning difficulties and dysmorphic features. Chromosomal analysis using fluorescence in situ hybridization (FISH) analysis showed a deletion of chromosome 22q11.2 and confirmed the diagnosis of DiGeorge syndrome. This case report demonstrates that DiGeorge syndrome should be considered while investigating hypocalcaemia and Hypoparathyroidism in adulthood as this syndrome has very important implications for health and future family planning for patients and their families.

Case Report

Our patient is a 40-year-old lady who presented to our department feeling unwell with fever and numbness in both hands. Past medical history showed recurrent urinary tract infections, rheumatoid arthritis, chronic eczema and epilepsy. She was taking Levetiracetam 500mg twice daily and Clobazam 5 mg twice daily for the epilepsy. She is also known to have learning difficulties. Mild hypocalcaemia was documented few years back in a previous admission in other hospitals, but the cause was unclear. On admission, she was hemodynamically stable with mild facial dysmorphism, and positive Trousseau's and Chvostek's signs.

Blood tests showed a low corrected calcium 1.5 mmol/L (NR 2.25-2.5 mmol/L), high C-reactive protein, Leukocytosis, and 3.0 mmol/L serum potassium level (NR 3.5-5.0 mmol/L). Other routine blood tests were normal. Further investigations showed low Serum parathyroid hormone levels, normal magnesium levels and normal TSH level. A CT scan of the brain was unremarkable. Electrocardiogram showed QT prolongation (with QTc of 520 ms). The diagnosis of hypoparathyroidism and urinary tract infection was established and the patient was treated with antibiotics to cover urinary tract infection and calcium supplements for hypocalcaemia. The patient symptoms improved significantly and was discharged on calcium supplements and Calcitriol (Rocaltrol 0.25 mcg) with a calcium level of 2.1 mmol/L. The presence of hypoparathyroidism in association with learning difficulties, eczema and epilepsy prompted chromosomal analysis for DiGeorge syndrome. The microdeletion of chromosome 22q11.2 was confirmed by FISH (fluorescent in situ hybridization) analysis. Cardiac echo examination demonstrated no abnormalities and abdominal ultrasound examination showed no renal abnormalities. The patient was offered Genetic counselling together with her family.

Discussion:

DiGeorge syndrome is a well-known genetic disorder with a prevalence of 1:4000 live births¹. It was initially described by Angelo DiGeorge a physician and paediatric endocrinologist in 1968 ². DiGeorge is a developmental defect caused by a microdeletion of chromosome 22q11.2; it is also known as velocardiofacial syndrome or CATCH 22 syndrome to describe the classical features of this syndrome (C-Congenital heart disease, A-Abnormal facies, T-Thymus hypoplasia, C-Cleft Palate and H- Hypocalcaemia due to Hypoparathyroidism. Autoimmune disorders, skeletal defects, renal abnormalities, psychiatric and behavioural disorders are also associated with this syndrome.

DiGeorge is an autosomal dominant syndrome but the majority of patients have de novo mutations caused mainly by the microdeletion of chromosome 22q11.2 which leads to developmental disorders such as the failure of development of pharyngeal pouch system ^{3, 4}. These developmental disorders are the main cause of the classic features and presentation of DiGeorge syndrome such as congenital heart diseases, hypoplasia of the parathyroid glands and thymus, congenital immune deficiency and renal abnormalities⁵.

Congenital Conotruncal cardiac defects that involve truncoaortic sac can present in 70% patients with DiGeorge Syndrome. The most common cardiac anomalies are interrupted aortic arch, Tetralogy of Fallot, Atrial septal defect and ventricular septal defects ^{4, 5, 6}.

Hypocalcaemia is due to hypoparathyroidism and is present in about 60 % of patients ^{5,7}. Hypocalcaemia is a strong predictor of DiGeorge syndrome if it is associated with other clinical features such as cardiac defect and immunodeficiency. Hypocalcaemia commonly presents as muscle cramps,

numbness, tetany, focal or generalized seizures, prolong QT and hypotension.

Immunodeficiency is rare in adults and but it may present in up to 70-80% of the children with DiGeorge syndrome. Immunodeficiency occurs because of the low T cell count due to thymus hypoplasia. The function of T cells is however, usually preserved. Patients with immunodeficiency may have recurrent viral chest infection, systemic fungal infections frequent bacterial infections § .

The characteristic facies of DiGeorge include long face, narrow palpebral fissures, broad nasal bridge, micrognathia and asymmetrical crying face.

Psychiatric disorders have been reported with 22q11.2 deletion syndromes such as schizophrenia, bipolar disorder, anxiety and affective disorders.

Other conditions that may be associated with DiGeorge are atopic disorders (asthma and eczema) rheumatoid arthritis, autoimmune thyroiditis, renal abnormalities (such as multicystic kidneys and Vesicoureteral reflux).

Conclusion:

Due to the variety of symptoms and the de novo mutations, DiGeorge Syndrome should be considered in adults presenting with hypocalcaemia due to hypoparathyroidism even in the absence of the classical features. The syndrome has significant health implications, and confirming the diagnosis is important for future family planning.

Competing Interests

None declared

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Chest pain and syncope in a middle-aged man

Deacon Zhao Jun Lee, Karan Saraf and Paul Sheridan

Abstract

A 46 year old man presented to the Emergency Department with chest pain and collapse with loss of consciousness. The history, examination and investigation findings are detailed below followed by five questions surrounding the pathophysiology, diagnosis and management of the condition.

Keywords: Brugada syndrome, channelopathy, sudden cardiac death, ventricular tachycardia, ventricular fibrillation, risk stratification, internal cardioverter defibrillator

Case history

A 46 year-old man presented to the Emergency department with chest pain and collapse, associated with loss of consciousness lasting several minutes. He had no significant past medical history and he had no risk factors for coronary artery disease. However, he did note a similar episode of collapse and loss of consciousness one year prior for which he did not seek medical attention. There was no known family history of heart disease or sudden death.

On examination he was haemodynamically stable with a blood pressure of 130/80 mmHg and heart rate of 85 beats per minute. Jugular venous pressure was measured at 2cm above the sternal angle and heart sounds were normal with no added sounds. His oxygen saturation was 98% on air and chest was clear to auscultation. Chest X-Ray demonstrated clear lung fields and laboratory investigations, including electrolytes and cardiac troponin T were within normal limits. Echocardiography showed a structurally normal heart. Figure 1 shows his 12-lead electrocardiogram (ECG) on admission.



Figure 1 – 12 lead ECG on admission

Questions

- 1. What is the likely diagnosis based on the clinical presentation and ECG findings?
- 2. What life-threatening arrhythmias can arise from this condition?

- 3. What is the pathophysiology of this condition?
- 4. How is the diagnosis of this condition made?
- 5. What treatment options are available for patients with this condition?

Answers

1. What is the likely diagnosis based on the clinical presentation and ECG findings?

Short answer

Brugada syndrome

Long answer

The ECG shows coved Type 1 ST segment elevation in keeping with a diagnosis of Brugada syndrome.

First described in 1992, Brugada syndrome is a primary cardiac electrical disease or channelopathy that is accompanied a structurally normal heart and carries an association with sudden cardiac death¹.

Presentation often occurs in the third or fourth decades of life, with a male preponderance of 8:1, however sudden cardiac death due to Brugada syndrome has also been seen in patients at the extremes of age. It is estimated that it accounts for up to 20% of all sudden cardiac deaths in patients without structural heart disease, ischaemia or electrolyte abnormalities². It is most commonly seen in south-east Asia, especially Thailand, where its incidence is around 1%, and is much less common in western countries¹.

2. What life-threatening arrhythmias can arise from this condition?

Short answer

Brugada syndrome is associated with increased risk of ventricular tachycardia (VT), often polymorphic, and ventricular fibrillation (VF).

Long answer

Brugada syndrome often manifests clinically in the form of syncope or sudden cardiac death. It is most commonly associated with polymorphic VT and VF, which may or may not terminate spontaneously. There are a large proportion of patients with Brugada syndrome who never experience any symptoms and indeed, may not ever even be identified as having the condition, unless they are found to have incidental ECG abnormalities as part of routine medical testing or are under investigation for another problem³.

3. What is the pathophysiology of this condition?

Short answer

Brugada syndrome is understood as a genetic cardiac channelopathy, a disorder produced by the dysfunction of a cardiac ion channel participating in the action potential which can result in electrical change favouring the development of arrhythmias. Inheritance of the condition occurs via an autosomal dominant mode of transmission with incomplete penetrance⁴.

Long answer

Brugada syndrome is a genetic disorder, with a loss-of-function mutation of the SCN5A gene implicated in about 30% of sufferers. This gene codes for the α -subunit of the cardiac sodium channel. Other mutations of sodium and calcium channels have also been found. In inherited cases, the gene is passed in an autosomal dominant fashion, though sporadic mutations are also seen.

There is increased susceptibility to ventricular arrhythmias, because of altered depolarisation within the right ventricle. In SCN5A mutations, the defect in sodium channels leads to decrease in the sodium current and a shortening of the cardiac action potential by blunting phase 0 depolarisation. Potassium channels are also affected, with an increased number of transient outward potassium channel currents. This imbalance in the myocytes between sodium and potassium concentrations means the overall effect is to shorten the refractory period, making the myocytes more prone to re-entrant circuits, leading to the development of VT and degeneration to VF^{2,5}.

4. How is the diagnosis of this condition made?

Short answer

Brugada syndrome is characterised by electrocardiographic changes demonstrating coved ST segment elevation in the right precordial leads.

Long answer

Electrocardiographic abnormalities constitute the hallmark of Brugada syndrome. There are three different ECG patterns and in all three types, the ECG shows ≥2mm J point (junction between the termination of QRS complex and beginning of ST segment) elevation and a characteristically shaped ST segment in the right precordial leads⁶.

Type I has a 'coved' pattern ST segment elevation ≥2mm, with a descending terminal portion in at least one right precordial lead

Type II has a 'saddle-back' ST segment elevation ≥1mm and has a high elevation in its initial portion.

Type III has either coved or saddleback ST elevation but is less accentuated than types I or II (<1mm).

Although all the 3 patterns can be present in patients with Brugada syndrome, only the presence of a type-1 ECG pattern defines the diagnosis of the condition^{2,7}. The patterns for type II or III are not diagnostic, and carrying out a Class I antiarrythmic drug (AAD) test to confirm the diagnosis is recommended. This can be done with AADs such as ajmaline, flecainide or procainamide, though currently ajmaline is preferred due to its higher sensitivity in revealing Brugada type ECG changes¹.

It is worth noting that the resting ECG changes associated with Brugada syndrome (in particular type I) are often transient, and therefore, in someone in whom the diagnosis is suspected, an AAD test may be indicated even if there are no resting spontaneous ECG abnormalities evident⁶.

Differential diagnoses of Brugada syndrome must be approached with care as ST segment elevation is associated with a wide variety of benign and malignant pathophysiologic conditions³.

5. What treatment options are available for patients with this condition?

Short answer

Currently, the implantable cardioverter defibrillator is the only proven effective treatment in the prevention of sudden cardiac death.

Long answer

Management of Brugada syndrome is focused on risk stratification of patients to prevent arrhythmic death in high risk individuals. ICD implantation can prevent sudden cardiac death in these groups¹. Newer devices are now also being used, including the subcutaneous ICD which is implanted in a subcutaneous pocket and does not require any endovascular leads in the heart or access to the central venous circulation⁸.

Pharmacological options are focused on rebalancing the ion channel current active during the early phases of the epicardial action potential in the right ventricle^{3,9}. Some studies have evaluated the role of quinidine in the treatment of Brugada syndrome and found it to be effective in preventing polymorphic VT and VF in this condition¹⁰. Quinidine has also been proposed as an alternative to ICD implantation in children and infants too young to receive an ICD^{11,12}.

Data relative to the use of cryosurgical treatments or ablation therapy in Brugada syndrome are very limited at this point in time³.

Patient outcome

The patient underwent successful implantation of a subcutaneous cardioverter defibrillator. Figure 2 and Figure 3 show chest radiographs of the leadless device.

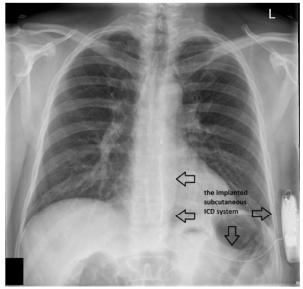


Figure 2 – Chest radiograph (PA view) showing the implanted subcutaneous ICD

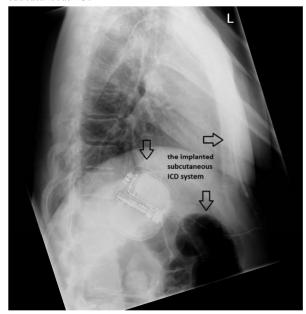


Figure 3 – Chest radiograph (lateral view) showing the implanted subcutaneous ICD

Competing Interests

None declared

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"Of Psychosis" - A Poem by Dr Javed Latoo

It all started at school when I was only sixteen Getting increasingly worried about my weight; All I wanted then was to get more slim and lean Fearing if I ate any food I may get overweight.

My whole life revolved around counting calories Making frantic efforts to avoid any fattening food; Surviving on just soup, toasts, coffee and berries The fear of being judged resulted a life in seclude.

Tireless dread of fatness and flabbiness of body Pushing to reduce weight by all means possible; Excessive exercises turning me thin like nobody Self induced vomiting made me feel horrible.

Things got even worse when I secretly started using Some water tablets and laxatives to look slimmer; My family got concerned when they saw bruising Finding me like a walking skeleton as I got leaner.

Thinning of hair, bad breath, dizziness, feeling cold Were just a few of the symptoms I experienced daily; Low moods and emaciation made me look very old Treading my days with a fat image in my head dizzily.

I lost all the features that made my body feminine Breasts, sexy hips, shining teeth and glowing skin; My worried family's apprehensions were genuine While I still dreamed of being a beautiful girl within.

It was impossible to convince me that I was not fat Until my nervous parents forced me to see a doctor; It has been a slow process of healing to arrive at The dawn where I saw the rebirth of their daughter.

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