

SITUS INVERSUS vis-à-vis SUDDEN DEATH

Dr. Adarsh Kumar, Senior Lecturer,

Dr. Virendar Pal Singh, Demonstrator,

Dr. Krishan Vij, Professor & Head,

Department of Forensic Medicine, Government Medical College, Sector 32, Chandigarh-160030.

ABSTRACT

Situs Inversus, a rare congenital anatomical abnormality although is not directly related to Sudden Death. However, 3-5% of people with Situs Inversus have associated functional heart defect & 25% have an underlying condition i.e. Primary Ciliary Dyskinesia presenting as Kartagener's Syndrome; are prone to Sudden Death.

Two cases of Situs Inversus on which medico-legal autopsy was conducted are reported. In first case, death was attributed to cardiac component (massive cardiomegaly, narrowing of coronaries and an old, healed apical scar) and in the second, due to respiratory complications (infection of lungs with areas of pus formation) with Blood Alcohol Concentration as 241.50 mg % ; being another significant finding.

Apart from discussing the various aspects of the condition, vital medico-legal issues varying from 'Negligence' to 'Assessment for Insurance purpose' are being discussed.

Key Words: Situs Inversus, Sudden Cardiac Death, Primary Ciliary Dyskinesia, Medicolegal Issues.

INTRODUCTION

Situs Inversus is a rare congenital anomaly in which there occurs reversal placement of various thoracic and abdominal organs to their normal anatomical location. In Situs Inversus totalis, which is a recessively inherited condition, there, occurs mirror image of the anatomic location of all the viscera while in Dextrocardia only the apex of the heart points to right and is present in the right hemithorax. Sometimes, when Situs Inversus is accompanied by sinusitis and bronchiectasis, it constitutes Kartagener Syndrome[1]. In all these conditions, person is apparently normal and average life spans have been reported. However, owing to susceptibility of these individuals to catch various infections, it contributes to morbidity and mortality in a person having pre-existing cardiovascular or respiratory lesions. Thus, the individuals having these lesions are prone to sudden death.

Sudden Death can be defined as one, which is not preceded or only preceded for a short time by morbid symptoms[2]. The WHO defines it as someone who dies within 24 hours of appearance of symptoms while currently it is defined as "Unexpected Death occurring within one hour of

new symptoms" [3]. The incidence of Sudden Death is about 10% of all causes of death and of these, almost half are due to cardiovascular causes and 15-23% is due to respiratory involvement[2,4]. Before the age of 35 years, the congenital and hereditary cardiovascular diseases are main causative factor for Sudden Death[3].

Sudden Cardiac Death (SCD) is defined as "death as a result of cardiac cause within 6 hours of onset of symptoms[5]". In 5-10% of SCD, hearts are apparently normal. Cardiomegaly is a frequent finding in cases that died suddenly from cardiovascular pathology[3]. 3-5% of people with SITUS INVERSUS have associated functional heart defect & 25% have primary ciliary dyskinesia presenting as Kartagener Syndrome.

Two such cases of SITUS INVERSUS on whom medico-legal autopsy was conducted owing to suspicious circumstances are being reported and

Case Report No. 1

An apparently healthy, 56 years old, male allegedly died suddenly on 1st January. Earlier, about 4 months back, he met with a road traffic accident, treated and later developed loss of memory for which he was diagnosed as having

chronic subdural hematoma on 31st December and further advised CT scan. However, he died the next day. On further investigation, it was revealed that in the month of June, he was clinically diagnosed as a case of Situs Inversus with Dextrocardia with global hypokinesia, Left Ventricular systolic and Left Ventricular diastolic dysfunction with hypertension.

Autopsy findings

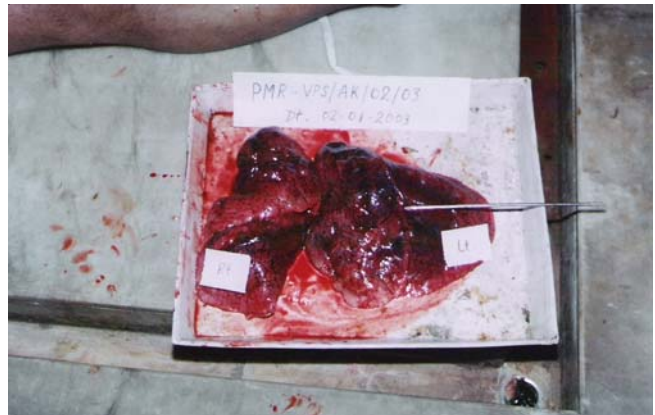
Average built and nourished dead body of an old male without any marks of external injury. On opening the body cavities, right lung weighed 400 gms and had two lobes while left lung had three lobes and weighed 550 gms. Heart was present on right side of chest cavity with apex pointing towards right side i.e. dextrocardia (photograph No. 1 & 2). The heart weighed 740 gms and had measurements as 17x14x7.5 cms (Photograph No. 3). Stomach was present on right side and contained 50 cc of whitish fluid without any suspicious smell and having normal mucosa. Liver and caecum were present on left side of abdominal cavity while spleen was present on right side.

Photograph 1: Showing reverse anatomical placement of various viscera. Heart present on the right side and apex pointing towards right side. Liver is present on left side.



On histopathological examination of heart, right and left ventricular walls measured 0.3cm and 0.8 cm respectively. A grey white area of 0.5x0.4cm was found at apex of heart corresponding to old, healed myocardial infarct. Cut section of coronaries showed atheromatous plaque causing narrowing of lumen. (Right coronary artery - 50%, Left anterior descending - 50%, Left circumflex - 25-30%). Chronic venous congestion was found in both liver and lungs, while no pathological change was found in brain.

Photograph 2: Right lung showing two lobes instead of three. Left lung showing three lobes instead of two.



Photograph 3: Enlarged heart (cardiomegaly) weight 740 grams. Measuring 17 x 14 x 7.5 cms.



Case Report No. 2

A middle-aged rickshaw puller was found dead under mysterious circumstances in his hut (jhuggi). Alleged history as given by nephew of the deceased was: the deceased, a known smoker & alcoholic, married but issueless; was seen alive last on 9th January, 1999 at about 9 P.M. He used to carry passengers to and from a lodge on his rickshaw and meet his nephew and other relatives in night after finishing this work. On the night of 10th January, he did not meet them. In the morning of 11th January, when his nephew went to see at his residence, he saw the door of jhuggi was not locked. As he pushed the door, it opened and he saw a dead body lying on cot with disfigured face. With the help of clothing, body structure and configuration, he recognized the body as of his uncle. He informed other relatives and police and

it was alleged that his uncle has been killed by his neighbour due to an old enmity. Also, he narrated an incidence of his uncle having a scuffle with the same neighbour 3-4 years back in which he suffered a fracture of right arm. Further, he correlated the statement of neighbour when that same neighbour threatened to eliminate his uncle, by killing and throwing against dogs. Autopsy could only be conducted after 3 days of recovery of dead body due to political intervention, owing to sensitive nature and constitution of 'Board of Doctors' for conducting postmortem examination.

Autopsy Findings

Average built and poorly nourished dead body of a middle aged male having put on a lot of clothing like shirt, jersey, pullover and covered with quilt and blanket. A 17x15 cm, U-shaped tissue defect with irregular margins involving front of neck and upper part of chest and without any vital reaction. Face was devoid of skin and underlying tissue, had crenated margins but without any evidence of antemortem hemorrhage (Photograph 4). Trachea absent, all fingernails showed bluish discoloration and body was in a state of advanced decomposition.

Photograph 4: Tissue defect in form of crenated margins evident over forehead without any signs of antemortem hemorrhage.



Entire body was subjected for radiological examination. Chest x-ray revealed the heart shadow and gas shadow of fundus of stomach on right side, but it was reported as normal by radiologist. The 'marker' for left side was not properly placed and in fact, it was placed almost in

the middle (Roentgenogram). On opening the chest cavity, the heart was present on right side but apex pointed towards left. An anomalous placement of aorta in form of origin i.e. anterior to pulmonary trunk was observed. Left lung had three lobes while right lung had two lobes and on cut section, foci of pus were present in both lungs. All other abdominal viscera were placed opposite to their normal anatomical location. Toxicological examination revealed a Blood Alcohol Concentration of 241.50 mg% while autolytic changes were found on histopathological examination of various viscera.

Photograph 5: X-ray chest AP view. Heart shadow on right side, gas shadow of stomach on right side. Note the presence of 'marker', which is almost in the middle.



DISCUSSION

The exact incidence of Situs Inversus is not known because persons remain asymptomatic; however, an incidence of 6-8 per 1000 live births is reported for congenital anomalies of heart⁶. The incidence of Kartagener Syndrome is about 1:20,000. 3-5% of people with Situs Inversus have associated functional heart defect, which is higher than rate of heart defect in general, population. Although Situs Inversus alone does not increase the risk of cardiovascular disease but in cases with associated cardiac anomaly, it may lead to Sudden Death.

In case report I, the weight of heart, which was taken after removing, blood and clots from chambers, was 740 gms. According to Reddy, the persons having their heart weight in excess of 420 gms, are prone to Sudden Death^[4]. Durigaon in

his study on 77 cases has stated that persons dying due to cardiovascular disease had a mean heart weight as 452gms[3]. There is a definite relationship between Body Mass i.e. both body height, weight, and heart mass and must be considered before labeling any heart as abnormal. Hitosugi et al has devised a simplified normal heart weight scale after taking into consideration Body Mass Index⁷. Kitzman et al devised formula for estimating heart mass from body mass based on sex and found it to be a better predictor of normal heart mass than either body surface area or body height⁸. Also the individuals with large heart weight are at risk for developing a dysarrhythmia and hence die unexpectedly^[8]. Heart weight in normal subject is also said to increase with age and physical activity, hypertension and alcoholism as well as cocaine abuse [8,9].

In the instant case, the body was of an average built and nourished old male, having a length of 5'3"; heart weight of 740 gms is definitely higher than normal expected weight. In addition, various coronary arteries showed 30-50% narrowing of their lumen, which collectively had their affect over the heart.

More than 75% stenosis of at least one segment of a major epicardial coronary artery without any other cardiac lesion can cause sudden death^[3]. In older adults i.e. age >45 yrs, 80-90% of sudden cardiac death have significant coronary artery disease while it is associated with 58-70% of sudden cardiac death in young adult population⁵. Congenital conditions comprise about 14% of structurally abnormal heart and next in occurrence to coronary artery disease i.e. about 65% cases^[5]. Congenital and hereditary cardiovascular diseases are main etiologies of Sudden Death before age of 35 years^[3]. Both obesity and epilepsy are co-morbid conditions in sudden cardiac death with structurally normal hearts.

No evidence of any gross or microscopic finding in brain was found which could substantiate the clinical diagnosis of subdural hemorrhage and supplemented the charge of Road Traffic Accident as cause for sudden death of this individual. Thus, due to presence of abovementioned findings with diagnosed hypertension during lifetime, it was opined that person was prone for Sudden Death and declared as a case of Sudden Cardiac Death.

In case Report 2, the external findings were declared as Postmortem Injuries. The presence of pus foci in lungs on gross examination favored the diagnosis of bronchiectasis although no histological comment was possible due to autolysis of viscera including lungs. As reported by Kartagener there occur Situs Inversus with chronic otitis media, chronic sinusitis and abnormalities leading to bronchiectasis due to loss of cilia in various cells leading to abnormal muco-ciliary clearance from middle ears, sinus cavities and airways. It is also seen in men with infertility secondary to sperm immobility due to same reason. Since blood alcohol concentration was 241.50 mg%, the person was prone to develop aspiration; also because of absence of ciliary movements in such a case. On further investigation, it was revealed that deceased was childless thereby favoring the diagnosis of Kartagener Syndrome. Thus in the absence of any antemortem traumatic pathology, taking due consideration of all findings; cause of death was opined as respiratory complications in presence of alcohol in a case of Situs Inversus/ Kartagener Syndrome.

CONCLUSION

If any case of Situs Inversus is encountered at autopsy, the family of deceased should be counseled and first-degree relatives may be advised by autopsy surgeon to undergo a possible screening, as they are prone to have various cardiac abnormalities^[3].

If it is diagnosed during lifetime, as these persons are extra-susceptible to catch various respiratory infections; the dangers of smoking and exposure to industrial fumes should be explained by treating physician. An appropriate vocational guidance may also be supplied to them.

Failure to diagnose these cases by Physicians, Anesthetists and Radiologists may lead to charge of negligence against them^[10]. The inattention to proper labeling of side i.e. Right or Left on X-ray films can lead to non-recognition or missed diagnosis of Situs Inversus. If the condition is missed by Anesthetist during pre-anesthetic check-up, surgical mishaps may result.

Situs Inversus with cardiac abnormality or respiratory complications decreases the life span and thus increases risk for sudden death. This

requires focusing while assessing these individuals for insurance purposes.

Acknowledgement

Authors wish to accord their sincere appreciation to Ms. Rupinder Kaur for typing this manuscript. Thanks are also extended to department of Pathology, GMCH-32, Chandigarh for providing their comments on various histopathological findings.

References

1. Kobzik L. The Lung. In Robbin's Pathologic Basis of Disease ed. by Cotran, Kumar and Collins. Pub. by W.B. Saunders Co., Philadelphia, In India by Harcourt India Pvt. Ltd., New Delhi. 6th Ed. 2000: p.716.
2. K Adarsh. Sudden Death due to Acute Pancreatitis. Journal of Karnataka Medicolegal Society, 2001; 10 (1): 23-25.
3. Grandmaison GLDL, Durigon M. Sudden Adult Death: A Medicolegal series of 77 cases between 1995 & 2000. Medicine, Science and Law, 2002; 42 (3): 225-32.
4. Narayan Reddy KS. Death and its cause. In the Essentials of Forensic Medicine & Toxicology. Pub. by K. Sugana Devi, Hyderabad, 22nd Ed., 2003 : p. 417.
5. Chugh SS, Kelly KL, Titus JL: Sudden Cardiac Death with apparently normal heart. Circulation, 2000; 102 : 649-654
6. Schoen FJ. The Heart. In Robbin's Pathologic Basis of Disease ed. by Cotran, Kumar and Collins. Pub. by WB. Saunders Co., Philadelphia, In India by Harcourt India Pvt. Ltd., New Delhi 6th Ed. 2000: p.592.
7. Hitosugi M, Takatsu A, Kinugasa Y and Takao H: Estimation of normal heart weight in Japanese subjects: development of a simplified normal heart weight scale. Legal Medicine, 1999; (1): 80-85.
8. Davis GG, McGwin JG. Comparison of Heart Mass in seizure patients dying of sudden unexplained death in epilepsy to Sudden Death due to some other cause. Am J. of Forensic Medicine & Pathology, 2004; 25(1):23-28.
9. Hangartner JRW, Marley NJ, Whitehead A, Thomas AC and Davies MJ. The assessment of cardiac hypertrophy at autopsy. Histopathology, 1985; 9: 1295-1306.
10. Deshmukh R and Shah VR. Situs Inversus, Medicolegal Approach JFMT, 1994 Vol.13, 3 & 4 p. 33-34.

INSTRUCTIONS TO AUTHORS

CONTRIBUTIONS:

Unpublished original manuscripts, written in English should be sent to:
Dr. R.K.Gorea, Editor, JIAFM

THE PUBLICATION PARTICULARS:

The JIAFM is the official publication of the Indian Academy of Forensic Medicine, Published quarterly (Jan, April, July, Oct.) from 1991.

THE CONTENTS OF THE JOURNAL:

The journal accepts a range of articles of interest, under several feature sections as follows:

- Original Papers: Includes conventional observational and experimental research.
- Commentary: Intended for Reviews, Case Reports, Preliminary Report and Scientific Correspondences.

Letter to the Editor:

Designed to be an avenue for dialogue between the authors of the papers published in the journal and the readers restricted to the options expressing reviews, criticisms etc. It could also publish letters on behalf of the current affairs in the field of Forensic medicine in the country.

Editorial:

Intended as a platform for the Editor-in-Chief and for others with a keen interest in forensic medicine that wished to comment on the current affairs.

Special Features:

In the History of Indian Forensic Medicine, Book Review, Abstracts, Announcement etc, which appear frequently, but not necessarily in every issue.

News and Notes:

Intended for providing information of members and activities of the Academy and its Chapters of State level / other such other organizations affiliated to the Academy. May appear frequently and not in every issue.

Preparing a Manuscript for Submission to a Biomedical Journal:

General Principles: The text of observational and experimental articles is divided into sections with the headings Introduction, Methods, Results, and Discussion. This so-called "IMRAD" structure is not simply an arbitrary publication format, but rather a direct reflection of the process of scientific discovery. Other types of articles, such as case reports, reviews, and editorials, are likely to need other formats. Double spacing of all portions of the manuscript - including the Title Page, Abstract, Key Words, Introduction, Methods, Results, Discussions, Conclusion, Acknowledgements, References, Tables, Figures and Legends - and generous margins with numbering all of the pages of the manuscript consecutively, beginning with the title page. Limits specified are Original Papers and Review Papers: 3000 words. Special Features and News & Notes 500 words. All other sections 1500 words. Submit ONE Original Typed copy in 8" x 11" Bond paper and TWO carbon copies on typing paper along with THREE sets of illustrations / Figures and Tables.

Title Page: The title page should carry the following information:

1. The Title of the Paper.
2. Authors' names and institutional affiliations with each author's highest academic degree.
3. The name of the department(s) and institution(s) to which the work should be attributed.
4. Disclaimers, if any and any details about the grants for the research.
5. Corresponding authors. The name, mailing address, telephone numbers, and e-mail address of the author responsible for correspondence about the manuscript.
6. The number of figures and tables.

Mention clearly to which feature section the manuscript is sent for.

Abstract and Key Words: An abstract not exceeding 100 words should follow the title page. The abstract should provide the context or background for the study and should state the study's purposes, basic procedures (selection of study subjects or laboratory animals, observational and analytical methods), main findings (giving specific effect sizes and their statistical significance, if possible), and principal conclusions. It should emphasize new and important aspects of the study or observations. Following the abstract, 3 to 10 key words or short phrases that capture the main topics of the article, will assist indexers in cross-indexing the article and may be published with the abstract. Terms from the Medical Subject Headings (MeSH) list of Index Medicus should be used.

Introduction: Provide a context or background for the study (i.e., the nature of the problem and its significance). State the specific purpose or research objective of, or hypothesis tested by, the study or observation; the research objective is often more sharply focused when stated as a question. Both the main and secondary objectives should be made clear, and any pre-specified subgroup analyses should be described. Give only strictly pertinent references and do not include data or conclusions from the work being reported.

Methods: The Methods section should include only information that was available at the time the plan or protocol for the study was written; all information obtained during the conduct of the study belongs in the Results section. Describe your selection of the observational or experimental participants (patients or laboratory animals, including controls) clearly, including eligibility and exclusion criteria and a description of the source population. Identify the methods, apparatus (give the manufacturer's name and address in parentheses), and procedures in sufficient detail to allow other workers to reproduce the results. Give references to established methods, including statistical methods; provide references and brief descriptions for methods that have been published but are not well known; describe new or substantially modified methods, give reasons for using them, and evaluate their limitations. Identify precisely all drugs and chemicals used.

Statistics: Describe statistical methods with enough detail to enable a knowledgeable reader with access to the original data to verify the reported results. When possible, quantify findings and present them with appropriate indicators of measurement error or uncertainty (such

as confidence intervals). Avoid relying solely on statistical hypothesis testing, such as the use of P values, which fails to convey important information about effect size. Define statistical terms, abbreviations, and most symbols. Specify the computer software used.

Results: Present your results in logical sequence in the text, tables, and illustrations, giving the main or most important findings first. Do not repeat in the text all the data in the tables or illustrations; emphasize or summarize only important observations. When data are summarized in the Results section, give numeric results not only as derivatives (for example, percentages) but also as the absolute numbers from which the derivatives were calculated, and specify the statistical methods used to analyze them. Use graphs as an alternative to tables with many entries; do not duplicate data in graphs and tables. Avoid non-technical uses of technical terms in statistics, such as “random”, “normal,” “significant,” “correlations,” and “sample.”

Discussion: Emphasize the new and important aspects of the study and the conclusions that follow from them. Do not repeat in detail data or other material given in the Introduction or the Results section. For experimental studies it is useful to begin the discussion by summarizing briefly the main findings, then explore possible mechanisms or explanations for these findings, compare and contrast the results with other relevant studies, state the limitations of the study, and explore the implications of the findings for future research and for clinical practice. Link the conclusions with the goals of the study but avoid unqualified statements and conclusions not adequately supported by the data. In particular, authors should avoid making statements on economic benefits and costs unless their manuscript includes the appropriate economic data and analyses. Avoid claiming priority and alluding to work that has not been completed. State new hypotheses when warranted, but clearly label them as such.

References: References to review articles can be an efficient way of guiding readers to a body of literature; review articles do not always reflect original work accurately. Small numbers of references to key original papers will often serve as well as more exhaustive lists. Avoid using abstracts as references. For the reference style and format the uniform requirements of the International Committee of Medical Journal Editors are used. Should be numbered singly in the order in which they are cited (using numerical in square brackets in the text and listed in numerical sequence on a separate sheet. Reference to journals, book and multi author volumes should accord with following examples.

1. Sivaloganathan S and Butt WP. A foot in the Yard. Med. Sci. Law, 1988; 28: 150-155.
2. Mukherji JB. Forensic Medicine and Toxicology. 1st Ed. Calcutta: Academic Publishers; 1981. p.72.

Personal Communications, Unpublished Papers etc. are not allowed as references.

Tables: Tables capture information concisely, and display it efficiently; they also provide information at any desired level of detail and precision. Including data in tables rather than text frequently makes it possible to reduce the length of the text. Type or print each table with double spacing on a separate sheet of paper. Number tables consecutively in the order of their first citation in the text and supply a brief title for each. Do not use internal horizontal or vertical lines. Authors should place explanatory matter in footnotes, not in the heading. Identify

statistical measures of variations, such as standard deviation and standard error of the mean. Be sure that each table is cited in the text.

Illustrations (Figures): Figures should be either professionally drawn and photographed, or submitted as photographic quality prints. In addition electronic files of figures in a format (e.g., JPEG or GIF) will produce high quality images. For x-ray films, scans, and other diagnostic images, as well as pictures of pathology specimens or photomicrographs, send sharp, glossy, black-and-white or color photographic prints, usually 127 x 173 mm (5 x 7 inches). Letters, numbers, and symbols on Figures should therefore be clear and even throughout, and of sufficient size that when reduced for publication each item will still be legible. Titles and detailed explanations belong in the legends, however, not on the illustrations themselves. Photomicrographs should have internal scale markers. Symbols, arrows, or letters used in photomicrographs should contrast with the background. If photographs of people are used, either the subjects must not be identifiable or whenever possible permission for publication should be obtained. Figures should be numbered consecutively according to the order in which they have been first cited in the text. Type or print out legends for illustrations using double spacing, starting on a separate page, with Arabic numerals corresponding to the illustrations. When symbols, arrows, numbers, or letters are used to identify parts of the illustrations, identify and explain each one clearly in the legend. Explain the internal scale and identify the method of staining in photomicrographs. Indicate Figure Number and an arrow pointing the top, on the backside of each photograph with a pencil lightly.

Units of Measurement: Measurements of length, height, weight, and volume should be reported in metric units (meter, kilogram, or liter) or their decimal multiples. Temperatures should be in degrees Celsius. Blood pressures should be in millimeters of mercury, unless other units are specifically required by the journal. Report laboratory information in both the local and International System of Units (SI).

Abbreviations and Symbols: Use only standard abbreviations; the use of non-standard abbreviations can be extremely confusing to readers. Avoid abbreviations in the title. The full term for which an abbreviation stands should precede its first use in the text unless it is a standard unit of measurement.

Sending the Manuscript to the Journal: Journal now accepts **electronic submission** of manuscripts, either on disk, as attachments to electronic mail (editor's email). Electronic submission saves time as well as postage costs, and allows the manuscript to be handled in electronic form throughout the editorial process. If a paper version of the manuscript is submitted, send the required number of copies of the manuscript and figures.

The letter should give any additional information that may be helpful to the editor, such as the type or format of article. If the manuscript has been submitted previously to another journal, it is helpful to include the comments.

The ICMJE has neither endorsed nor approved the contents of this reprint. The ICMJE periodically updates the Uniform Requirements, so this reprint prepared on may not accurately represent the current official version at <http://www.icmje.org/>