ORIGINAL ARTICLE

Awareness about Patterson syndrome among dental students

M. Dhakshinya,
Vishnu Priya Veeraraghavan,
R. Gayathri, S. Kavitha

Department of Biochemistry, Saveetha Dental College and Hospitals, Saveetha Institute of Medical and Technical Sciences, Saveetha University, Chennai, Tamil Nadu, India

J. Adv. Pharm. Technol. Res.

ABSTRACT

The aim is to create awareness about Patterson syndrome among dental students. Patterson-Stevenson-Fontaine syndrome is a very rare condition marked by irregular facial bone and tissue growth (mandibulofacial dysostosis) as well as limb abnormalities. A recessed jaw (retrognathism), cleft palate, and external ear defects are all possible symptoms of this disorder. A total of 112 undergraduate dental students participated in a longitudinal cross-sectional sample. To assess college students' awareness about Patterson syndrome, a self-administered, closed-ended questionnaire was developed and distributed. The only language allowed was English. The results were analyzed in SPSS software version 23. 10.71% of females and 14.29% of males were aware about Patterson syndrome. 32.14% of females and 38.39% of males were aware that Patterson syndrome was a rare adrenal disorder. We can conclude that very few of the population which was only 25% of the dental students were aware about Patterson syndrome and this survey helped in creating awareness about this syndrome.

Key words: Innovative technique, mandibulofacial deformities, novel method, Patterson syndrome, syndactyly

INTRODUCTION

Patterson-Stevenson-Fontaine syndrome is a very rare condition marked by irregular facial bone and tissue growth (mandibulofacial dysostosis) as well as limb abnormalities.[1] A recessed jaw (retrognathism), cleft palate, and external ear defects are all possible symptoms of this disorder.[2] The split-foot deformity is also seen in this syndrome, which is also known as ectrodactyly; hence, Patterson syndrome is the combination of these limb defects. Patterson-Stevenson-Fontaine syndrome is passed down through the generations in an autosomal dominant pattern.[3] A person's symptoms that are associated with the syndrome can be used to diagnose the disorder. The goal of treatment is to alleviate each person's symptoms.[4] Patterson-Stevenson-Fontaine syndrome is characterized by symptoms that begin at birth. Face characteristics and a split-foot deformity are among the signs. Mandibulofacial dysostosis is characterized by a receding jaw (retrognathism), a cleft palate, and changes in the external ear features.[5] Patterson-Stevenson-Fontaine syndrome affects some people with all of these symptoms, while others only have symptoms that affect their face or feet.[6] The causes of this syndrome are still a mystery to scientists. It may be caused by a particular genetic alteration (mutation), but the gene responsible has yet to be discovered. Since the syndrome's signs are present from birth, a mutation in a gene that governs the development of the face and feet is most likely to blame.[7]

Address for correspondence:
Dr. Vishnu Priya Veeraraghavan,
Department of Biochemistry, Saveetha Dental College and Hospitals, Saveetha Institute of Medical and Technical Sciences, Saveetha University, Chennai - 600 077, Tamil Nadu, India.
E-mail: drvishnupriyav@gmail.com

Submitted: 16-May-2022 **Published:** 30-Dec-2022
Accepted: 14-Oct-2022

Access this article online	
Quick Response Code:	Website: www.japtr.org
	DOI: 10.4103/japtr.japtr_382_22

How to cite this article: Dhakshinya M, Veeraraghavan VP, Gayathri R, Kavitha S. Awareness about Patterson syndrome among dental students. J Adv Pharm Technol Res 2022;13:S392-6.

Patterson-Stevenson-Fontaine syndrome is passed down through the generations in an autosomal dominant pattern. This means that a modification (mutation) in only one copy of the responsible gene in each cell is sufficient to trigger the syndrome's symptoms.[8] This syndrome has yet to be pinpointed as to what gene is responsible for the proper development of the face and feet. When an individual with Patterson syndrome has offspring, each offspring has a 50% (1 in 2) risk of inheriting the syndrome's gene change. Patterson-Stevenson-Fontaine syndrome, on the other hand, has been stated to have a lower penetrance rate.[9] This means that not everyone who has a disease-causing mutation will develop symptoms. As a result, this state has the potential to "skip" generations. When a person with the syndrome has a child who has the gene change but no symptoms of the syndrome, the child can develop symptoms of the syndrome when the person has more children. Patterson syndrome manifests itself in a variety of ways. This means that not everyone who is affected may experience the same effects, and some individuals may be more seriously affected than the others.[10,11] Extensive research experience and deep knowledge of our team helped to translate it into publications of high quality.[12-26] The aim of this survey is to assess the awareness of Patterson syndrome among the college students.

MATERIALS AND METHODS

A total of 112 dental undergraduates participated in a longitudinal cross-sectional sample. To assess college students.' awareness about Patterson syndrome, a self-administered, closed-ended questionnaire was developed and distributed. The only language allowed was English. The statistical analysis was done by SPSS software (IBM, India). The association between the variables in comparison with gender was assessed by Chi-square test and Pearson correlation analysis. $P < 0.05$ is considered to be significant statistically.

RESULTS AND DISCUSSION

The results showed that 10.71% of females and 14.29% of males were aware about Patterson syndrome [Figure 1]. 32.14% of females and 38.39% of males were aware that Patterson syndrome was a rare adrenal disorder [Figure 2]. Only 7.14% of females and 14.29% of males were aware that Retrognatism, cleft palate and anomalies of the external ear are symptoms of Patterson syndrome [Figure 3]. Fifty percent of the participants have replied that diagnosis of Patterson syndrome is based on symptoms [Figure 4]. 43.75% of the students have replied that surgery for this syndrome is possible only in presence of abnormalities like cleft palate or syndactyly [Figure 5]. Only 44.64% of the participants were aware that Patterson syndrome is an autosomal dominant disorder [Figure 6]. 54.47% of the participants replied that Patterson syndrome has mandibulofacial dysostosis [Figure 7]. 39.39% of the participants were aware that Patterson syndrome is also known as Leprechaunism [Figure 8]. There were 49.11% of female participants and 50.89 of male participants [Figure 9].

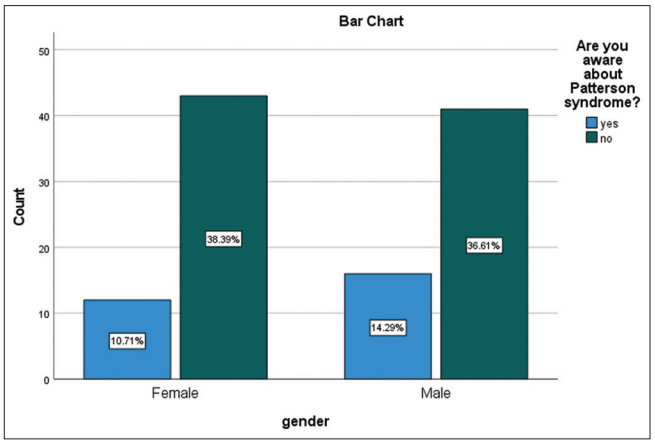

Figure 1: The graph shows the awareness of participants about Patterson syndrome. X-axis indicates gender and Y-axis indicates the responses. Blue indicates "yes" and green is "no." 10.71% of females and 14.29% males were aware whereas 38.39% of females and 36.61% of males were not aware. $P = 0.293$, (>0.05) hence, not significant statistically

Patterson syndrome is diagnosed when a person exhibits symptoms that are consistent with the syndrome. On the basis of ultrasound, it may be possible to diagnose the condition before a child is born (prenatally). However, since the condition is so uncommon, it is unlikely to be discovered unless a family member suspects it.[27] Patterson syndrome is treated by focusing on the individual's specific symptoms. Any of the symptoms of the condition, such as cleft palate and syndactyly, can be treatable with surgery. However, not all symptoms of the condition can be treated with surgery. Hearing aids may be required for people who have hearing loss. Since Patterson-Stevenson-Fontaine syndrome is so uncommon, the long-term outlook for those who are affected is unknown.[28] This syndrome has been linked to the birth of infants, and it does not appear to affect life expectancy. The etiology of Patterson syndrome is unknown, as discussed. However, it was observed that glycosaminoglycan secretion was abnormal qualitatively and quantitatively in some Patterson syndrome patients, but patterns of mucopolysaccharidoses are difficult to assess. Patterson patients usually have normal mucopolysaccharides observed in urine. The radiographic showings are distinct and unlikely to be mistaken for any other type of bone dysplasia.[29] While there are similarities to polyostotic fibrous dysplasia, hypophosphatasia, and certain types of metaphyseal chondrodysplasia on the surface. Because the overall picture and natural history

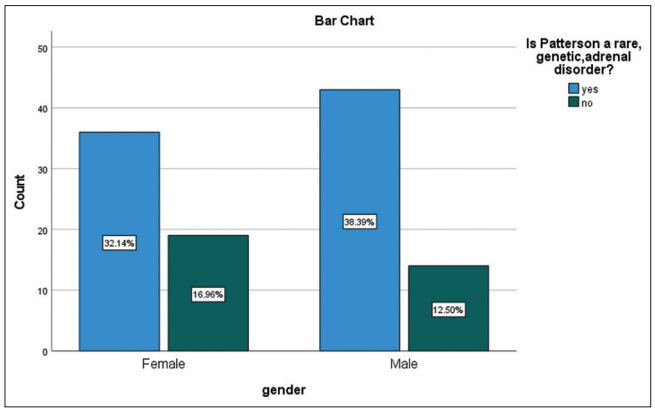

Figure 2: The graph represents whether the participants were aware that Patterson syndrome is a rare, genetic and adrenal disorder. X-axis indicates gender and Y-axis indicates the responses. Blue indicates "yes" and green is "no". 32.14% of females and 38.39% males were aware whereas 16.96% of females and 12.50% of males were not aware. $P = 0.171$, (>0.05) hence, not significant statistically

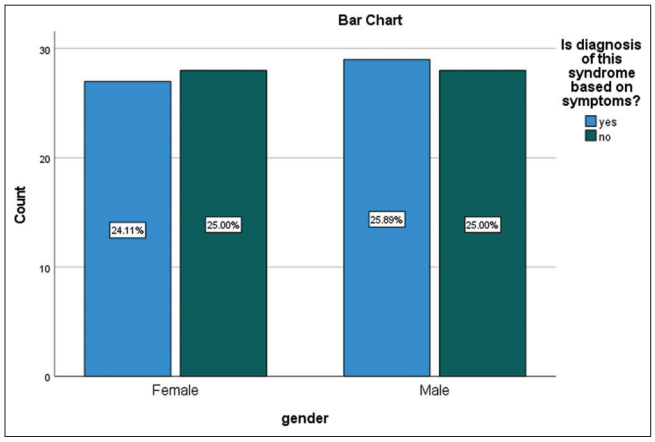

Figure 4: The graph represents whether the participants were aware that diagnosis of Patterson syndrome is based on symptoms. X-axis indicates gender and Y-axis indicates the responses. Blue indicates "yes" and green is "no". 24.11% of females and 25.89% males were aware whereas 25% of females and 25% of males were equally not aware. $P = 0.500$, (>0.05) hence, not significant statistically

of the condition are so dissimilar, there should be no difficulty in making a differential diagnosis.[30]

The survey was conducted only among dental students of Saveetha Dental College, so future surveys can be done, including other colleges from various parts of India, so we could assess the awareness of various hereditary disorders like Patterson syndrome. The survey could help in the early diagnosis and assessment of complications of Patterson syndrome through some oral manifestation which can, in turn, prevent more severe complications of this disease.

Limitation

The limitation of this study is smaller population and same geographic location.

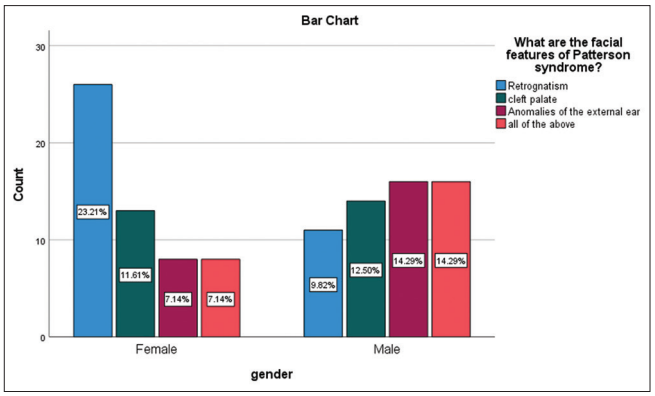

Figure 3: The graph represents whether the participants were aware about the facial features of Patterson syndrome. X-axis indicates gender and Y-axis indicates the responses. Blue indicates "Retrognatism", green indicates "cleft palate", red is "anomalies of the external ears" and orange represents "all of the above". 23.21% of females and 9.82% of males say it as Retrognathism, 11.61% of females and 12.50% of males have replied as cleft palate, 7.14% of females and 14.29% of males have replied as anomalies of the external ear. 7.14% of females and 14.29% of males have given a reply as all of the above. $P = 0.002$, (<0.05) hence, not significant statistically

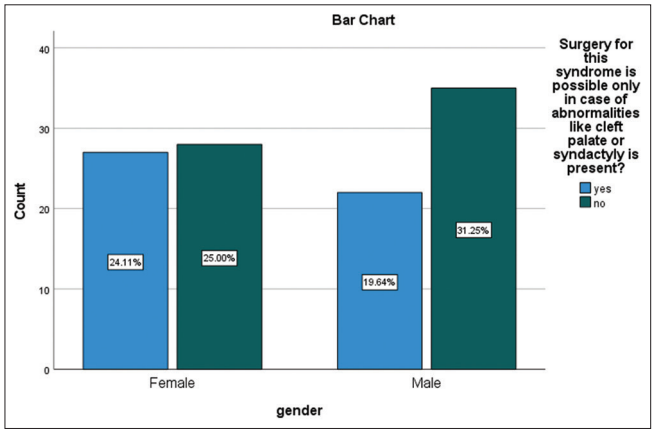

Figure 5: The graph represents whether the participants were aware that surgery for Patterson syndrome is possible only in case of abnormalities like cleft palate or syndactyly is present. X-axis indicates gender and Y-axis indicates the responses. Blue indicates "yes" and green is "no". 24.11% of females and 19.64% males were aware whereas 25% of females and 31.25% of males were not aware. $P = 0.117$, (>0.05) hence, not significant statistically

Future scope

Future studies should be conducted in a larger population and a wider range of geographically located people should be included.

CONCLUSION

Creating awareness plays a key role in providing the best treatment and early diagnosis of a disorder. The survey showed that the awareness of Patterson syndrome among the dental students was only average. Hence, this survey

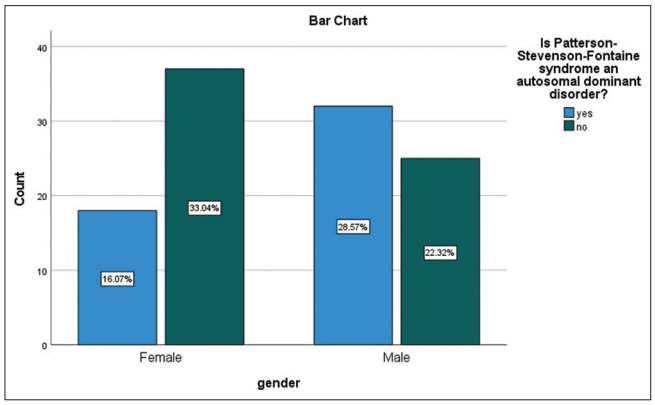

Figure 6: The graph represents whether the participants were aware that Patterson syndrome is an autosomal dominant disorder. X-axis indicates gender and Y-axis indicates the responses. Blue indicates "yes" and green is "no". 16.07% of females and 28.57% males were aware whereas 33.04% of females and 22.32% of males were not aware. $P = 0.010$, (<0.05) hence, significant statistically

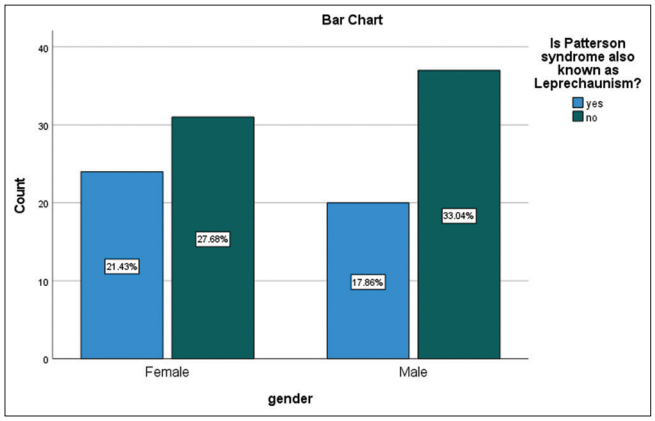

Figure 8: The graph represents whether the participants were aware that Patterson syndrome is also known as Leprechaunism. X-axis indicates gender and Y-axis indicates the responses. Blue indicates "yes" and green is "no". 21.43% of females and 17.86% males were aware whereas 27.68% of females and 33.04% of males were not aware. $P = 0.232$, (>0.05) hence, not significant statistically

helped in creating awareness about Patterson syndrome among dental students.

Acknowledgment

The authors would like to express their gratitude to Saveetha Dental University and Hospitals for assisting and for the successful completion of this task.

Financial support and sponsorship

The present project is supported by Saveetha Dental College, Saveetha Institute of Medical and Technical Science, Saveetha University, Dr. Murugesan Dental Clinic, Chennai.

Conflicts of interest

There are no conflicts of interest.

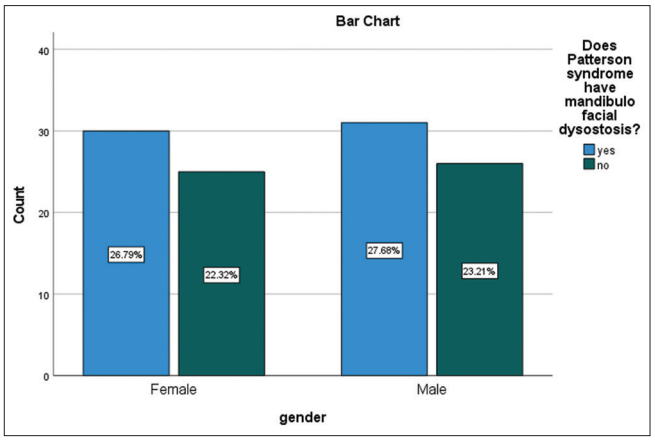

Figure 7: The graph represents whether the participants were aware that Patterson syndrome has any mandibulo facial dysostosis. X-axis denotes gender and Y-axis denotes the responses. Blue indicates "yes" and green indicates "no". 26.79% of females and 27.68% males were aware whereas 22.32% of females and 23.21% of males were not aware. $P = 0.569$, (>0.05) hence, not significant statistically

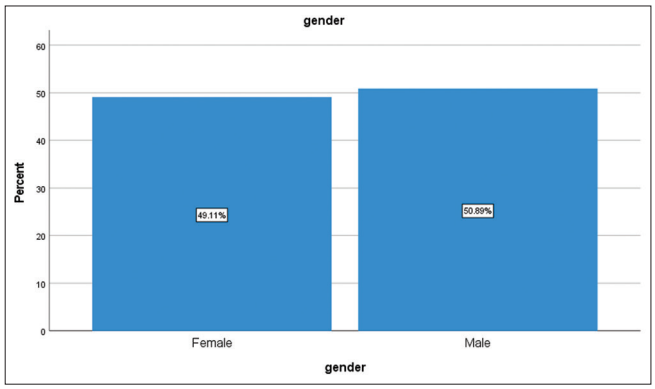

Figure 9: The graph represents the gender of the participants. X-axis denotes gender and Y-axis denotes the responses. 49.11% of females and 50.89% of male participants

REFERENCES

1. Keleszade E, Patterson M, Trangmar S, Guinan KJ, Costabile A. Clinical efficacy of brown seaweeds *Ascophyllum nodosum* and *Fucus vesiculosus* in the prevention or delay progression of the metabolic syndrome: A review of clinical trials. Molecules 2021;26:714.
2. Liu YA, Chijiwa C, Dunham CP, Jamieson DH, Solimano A, Schalkwyk JV, *et al.* Pathologic skull fracture in a near-term neonate with arthrochalasia type Ehlers-Danlos syndrome: A case report. Fetal Pediatr Pathol 2022;41:149-54.
3. Patterson Tichy AM, Bradley C. Unilateral exertional compartment syndrome in a pediatric competitive figure skater. Cureus 2019;11:e5611.
4. Patterson K, Kay J. Letter-by-letter reading: psychological descriptions of a neurological syndrome. Q J Exp Psychol A 1982;34:411-41.
5. Serson N. Dysostosis mandibulo facialis (maxillo-facial dysostosis). Plast Reconstr Surg 1958;21:237.
6. Wilson TG. A case of unilateral mandibulo-facial dysostosis associated with agenesis of the homolateral lung. J Laryngol Otol 1958;72:238-49.
7. Livak KJ, Little WA, Stack SL, Patterson TA. Polymorphisms in the

human DNA ligase I gene (LIG1) including a complex GT repeat. Mutat Res 1998;406:1-8.

8. Zlotina A, Melnik O, Fomicheva Y, Skitchenko R, Sergushichev A, Shagimardanova E, *et al.* A 300-kb microduplication of 7q36.3 in a patient with triphalangeal thumb-polysyndactyly syndrome combined with congenital heart disease and optic disc coloboma: A case report. BMC Med Genomics 2020;13:175.
9. Owen CI, Bowden R, Parker MJ, Patterson J, Patterson J, Price S, *et al.* Extending the phenotype associated with the CSNK2A1-related Okur-Chung syndrome-A clinical study of 11 individuals. Am J Med Genet A 2018;176:1108-14.
10. Chong JX, Yu JH, Lorentzen P, Park KM, Jamal SM, Tabor HK, *et al.* Gene discovery for Mendelian conditions via social networking: De novo variants in KDM1A cause developmental delay and distinctive facial features. Genet Med 2016;18:788-95.
11. Chong JX, McMillin MJ, Shively KM, Beck AE, Marvin CT, Armenteros JR, *et al.* De novo mutations in NALCN cause a syndrome characterized by congenital contractures of the limbs and face, hypotonia, and developmental delay. Am J Hum Genet 2015;96:462-73.
12. Nair M, Jeevanandan G, Vignesh R. Comparative evaluation of post-operative pain after pulpectomy with k-files, kedo-s files and mtwo files in deciduous molars-a randomized clinical trial. Braz Dent J 2018;21:411-7. Available from: https://bds.ict.unesp.br/index.php/cob/article/view/1617.
13. Kannan R, Thenmozhi MS. Morphometric study of styloid process and its clinical importance on Eagle's syndrome. Res J Phar Technol 2016;9:1137-9.
14. Samuel AR, Thenmozhi MS. Study of impaired vision due to Amblyopia. Pharm Res 2015;8:912-4. Available from: https://www.indianjournals.com/ijor.aspx?target=ijor: rjpt&volume=8&issue=7&article=019.
15. Viswanath A, Ramamurthy J, Dinesh SP, Srinivas A. Obstructive sleep apnea: Awakening the hidden truth. Niger J Clin Pract 2015;18:1-7.
16. Dinesh SP, Arun AV, Sundari KK, Samantha C, Ambika K. An indigenously designed apparatus for measuring orthodontic force. J Clin Diagn Res 2013;7:2623-6.
17. Varghese SS, Thomas H, Jayakumar ND, Sankari M, Lakshmanan R. Estimation of salivary tumor necrosis factor-alpha in chronic and aggressive periodontitis patients. Contemp Clin Dent 2015;6:S152-6.
18. Priyanka S, Kaarthikeyan G, Nadathur JD, Mohanraj A, Kavarthapu A. Detection of cytomegalovirus, Epstein-Barr virus, and torque teno virus in subgingival and atheromatous plaques of cardiac patients with chronic periodontitis. J Indian Soc Periodontol 2017;21:456-60.
19. Panda S, Jayakumar ND, Sankari M, Varghese SS, Kumar DS. Platelet rich fibrin and xenograft in treatment of intrabony defect. Contemp Clin Dent 2014;5:550-4.
20. Kamisetty SK, Verma JK, Arun, Sundari S, Chandrasekhar S, Kumar A. SBS versus inhouse recycling methods-an *in vitro* evaluation. J Clin Diagn Res 2015;9:ZC04-8.
21. Muthukrishnan A, Warnakulasuriya S. Oral health consequences of smokeless tobacco use. Indian J Med Res 2018;148:35-40.
22. Neelakantan P, Sharma S, Shemesh H, Wesselink PR. Influence of irrigation sequence on the adhesion of root canal sealers to dentin: A Fourier transform infrared spectroscopy and push-out bond strength analysis. J Endod 2015;41:1108-11.
23. Sahu D, Kannan GM, Vijayaraghavan R. Carbon black particle exhibits size dependent toxicity in human monocytes. Int J Inflam 2014;2014:827019.
24. Jose J, Ajitha P, Subbaiyan H. Different treatment modalities followed by dental practitioners for Ellis class 2 fracture – A questionnaire-based survey. Open Dent J 2020;14:59-65.
25. Uthrakumar R, Vesta C, Raj CJ, Krishnan S, Das SJ. Bulk crystal growth and characterization of non-linear optical bisthiourea zinc chloride single crystal by unidirectional growth method. Curr Appl Phys 2010;10:548-52.
26. Jain SV, Vijayakumar Jain S, Muthusekhar MR, Baig MF, Senthilnathan P, Loganathan S, *et al*. Evaluation of three-dimensional changes in pharyngeal airway following Isolated Lefort one osteotomy for the correction of vertical maxillary excess: A prospective study. J Maxillofac Oral Surg 2019;18:139-46.
27. Wilkie AO, Goodacre TE. Patterson-Stevenson-Fontaine syndrome: 30-year follow-up and clinical details of a further affected case. Am J Med Genet 1997;69:433-44.
28. David TJ, Webb BW, Gordon IR. The Patterson syndrome, leprechaunism, and pseudoleprechaunism. Journal of Medical Genetics 1981;18:294-8.
29. Latta RJ, Graham CB, Aase J, Scham SM, Smith DW. Larsen's syndrome: A skeletal dysplasia with multiple joint dislocations and unusual facies. The Journal of Pediatrics 1971;78:291-8.
30. Wang KH, Kupa J, Duffy KA, Kalish JM. Diagnosis and Management of Beckwith-Wiedemann Syndrome. Front Pediatr 2020;7:562. doi: 10.3389/fped.2019.00562.