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Presentation and non-surgical endodontic treatment of two patients with X-linked hypophosphatemia: a case report

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Abstract

Aim To describe two patients with X-linked hypophosphatemia presenting with spontaneous signs of pulpal necrosis in multiple intact teeth. The presentation and management are discussed, along with the diagnostic and endodontic treatment challenges

Summary Two young male patients with X-linked hypophosphatemia were referred to the Department of Restorative Dentistry at the Edinburgh Dental Institute for management of dental infection. Both patients were referred due to their unusual clinical presentation and abnormal root canal morphology. They subsequently presented on multiple occasions with pain or sinus tracts over a 3- and 5-year period whilst under care. Clinical examination revealed intact teeth with buccal swellings, draining buccal sinuses and negative responses to sensibility testing. Radiographic examination, generally consisting of periapical radiographs, revealed intact teeth with a range of unusual morphological features including large pulp chambers, wide canals, short roots and open apices, all with associated periapical radiolucencies. Due to the unusual morphology, some teeth required apexification with a mineral trioxide aggregate plug. Patient 1 underwent root canal treatment on teeth 21 and 43 over a 3-year period. Patient 2 underwent root canal treatment on 10 permanent teeth over a 5-year period. At follow-up, both patients were asymptomatic and clinically the teeth had no signs of infection or periapical inflammation. Radiographic examination confirmed complete resolution of the apical radiolucencies on 11 out of 12 teeth. Favourable outcomes have been demonstrated up to a follow-up of 4.5-years.

Key learning points

- Patients with X-linked hypophosphatemia may present with ‘spontaneous’ signs of pulp necrosis in multiple teeth in the absence of caries and trauma posing a diagnostic challenge.
- Abnormal morphological features, including wide canals and open apices, may present challenges during endodontic treatment.
- Outcomes demonstrate that an appropriate root canal treatment protocol, including the application of apexification procedures, may be implemented to successfully manage such cases.

Introduction

X-linked hypophosphatemia (XLH), also known as X-linked hypophosphatemic rickets or vitamin D-resistant rickets, is the most common form of hereditary rickets with a conservative prevalence estimate of less than 1 in 120,000 of the population in the United Kingdom (Foster *et al*. 2014, Hawley *et al.* 2020). XLH is an X-linked dominant genetic condition caused by a mutation inactivating the *PHEX* gene, responsible for phosphate metabolism. This leads to reduced phosphate reabsorption in the kidney causing hypophosphatemia as well as impaired mineralisation of bone and dentine (Nesbitt *et al*. 1999, Holm *et al.* 2001, Opsahl Vital *et al*. 2012, Foster *et al*. 2014, Beck-Nielsen *et al*. 2019). XLH manifests as skeletal abnormalities including reduced growth rate, short stature and bone deformity (Opsahl Vital *et al*. 2012, Beck-Nielsen *et al*. 2019).

Patients with XLH may present with dental abnormalities and complications both in the primary and permanent dentition. Published case reports and case series have consistently demonstrated spontaneous pulpal necrosis occurring in multiple teeth in the absence of caries or trauma (Goodman *et al.* 1998, Murayama *et al*. 2000, Alexander *et al*. 2001, Pereira *et al*. 2004, Batra *et al.* 2006, Douyere et al. 2009, Beltes & Zachou 2012, Soares et al. 2013, Lee *et al*. 2017, James & Roudsari 2019). Andersen *et al.* (2012) found patients with XLH had a characteristically high number of endodontically affected teeth with a mean of 4.2. Clinically, enamel defects have been reported, including hypoplasia and infractions (Goodman *et al.* 1998, Souza *et al*. 2010, Rabbani *et al*. 2012). Radiographically, a range of abnormal morphological features may be present, including enlarged pulp chambers and pulp horns, enlarged radicular canals, short roots and open apices (Hillmann & Geurtsen 1996, Goodman *et al.* 1998, Murayama *et al*. 2000, Alexander *et al*. 2001, Pereira *et al*. 2004, Souza *et al*. 2010, Lee *et al*. 2017). Histologically, enamel has occasionally been reported as thin and micro-cracks extending to the amelo-dentinal junction have also been identified (Douyere *et al.* 2009, Opsahl Vital *et al*. 2012). Dentine may be poorly mineralised with a widened predentine layer, lack of fusion of calcospherites and the presence of interglobular dentine (Abe *et al.* 1988, Seeto & Seow 1991, Hillmann & Geurtsen 1996, Zambrano *et al.* 2003, Chaussain-Miller *et al.* 2007, Opsahl Vital *et al*. 2012, Coyac *et al*. 2018). Primary teeth have occasionally been found to have extensions of the pulp horns to the amelo-dentinal junction, but no cases have detected this in permanent teeth (Goodman *et al.* 1998, Chaussain-Miller *et al.* 2007). The micro-cracks in enamel and exposed dentine through wear of the thin enamel may allow a pathway for bacteria to progress through the defective dentine and towards the prominent pulp horns resulting in 'spontaneous' pulp necrosis and subsequent development of infection within the root canal system. Histological examinations have confirmed defects within dentine to be filled with microorganisms (Hillmann & Geurtsen 1996).

Root canal treatment (RCT) appears to be the treatment of choice for the majority of permanent teeth with spontaneous pulpal necrosis (Murayama *et al*. 2000, Alexander *et al*. 2001, Pereira *et al*. 2004, Beltes & Zachou 2012, Lee *et al*. 2017). Despite the abnormal morphology, the majority of case reports used either a cold lateral condensation or continuous wave technique; however, radiographic follow-up was not always presented (Alexander *et al*. 2001, Beltes & Zachou 2012, Lee *et al*. 2017).

Report

Two young adult males with XLH attended following referral from their General Dental Practitioner (GDP). They presented with multiple spontaneous infections of endodontic origin and abnormal canal morphology. RCT was successfully completed in both patients. This case report was prepared according to the PRICE 2020 Guidelines (Nagendrababu *et al.* 2020) (Figure 1).

Case 1

A 16-year-old male was referred by their GDP due to a discharging sinus associated with tooth 21. The GDP was concerned regarding the “large and oddly shaped root canal”. At the time of initial assessment in May 2016, the patient was asymptomatic and had no specific complaints. Medically, there was a confirmed diagnosis of XLH and he was previously medicated with Alfacalcidol and Phosphate Sandoz. The patient’s dental history revealed recurrent dental infections in the primary dentition. The family history revealed a maternal link to XLH with associated episodes of dental infection and tooth loss.

Extra-oral examination was unremarkable. A comprehensive endodontic examination was performed including assessment of tenderness to percussion, tenderness to palpation, presence or absence of a sinus, mobility and periodontal pocket probing depth. This revealed that tooth 21 was unrestored with a buccal swelling, draining buccal sinus and grade 1 mobility. Tooth 21 gave a negative response to cold testing (BlueFreeze, Prodentis, Saint-Maur-des-Fossés, France). Radiographic examination, consisting of a periapical radiograph (FOCUS™ 3 intra-oral unit, Instrumentarium Dental, Tuusula, Finland) using standard settings (0.2 s exposure time, 70 kVp and 7 mA), was performed to assess the root canal morphology and determine the presence or absence of apical pathosis. The image was processed using the software Sirona SIDEXIS

(Dentsply Sirona, Charlotte, NC, USA) and viewed on a 21-inch screen using PACS (Picture Archiving and Communication System) software (Carestream Health, Rochester, NY, United States). This revealed unusual coronal anatomy with a wide root canal space and a periapical radiolucency (Figure 2a). Tooth 21 was diagnosed with pulp necrosis and a chronic apical abscess.

Verbal, informed and valid consent was obtained and RCT was commenced under rubber dam isolation at the end June 2016. The apical size of the canal was gauged at size 90 and manual dynamic activation with sodium hypochlorite (Chloraxid 5.25%, Cerkamed, Stalowa Wola, Poland) was performed. An inter-visit dressing of calcium hydroxide (Ultracal, Ultradent, Cologne, Germany) was placed and on review there was resolution of the sinus and no swelling. Canal filling was completed at the second visit in August 2016 with use of a mineral trioxide aggregate (MTA) (Angelus, Londrina, Brazil) plug and gutta-percha backfill (B & L Biotech, Fairfax, VA, USA) due to the open apex. The tooth was then restored with a double seal comprised of bulk-fill composite (SDR; Dentsply Sirona, Charlotte, NC, USA) followed by an enamel shade of a nanohybrid composite (Filtek™ Supreme; 3M ESPE AG, Seefeld, Germany) and a post-operative periapical radiograph revealed a satisfactory root filling (Figure 2b). An 8-month follow-up in April 2017 revealed an asymptomatic 21 which clinically had no associated signs of infection or periapical inflammation. Radiographically, there was evidence of a normal periodontal ligament space around the root (Figure 2c). The outcome was deemed to be favourable according to European Society of Endodontology Quality Guidelines and the patient was discharged (European Society of Endodontology 2006).

The patient was then re-referred in May 2019, 3 years following the initial referral, after development of a second spontaneous dental abscess associated with tooth 43 and an initial assessment was performed in June 2019. He initially complained of temperature sensitivity lasting a few weeks which had subsequently led to a severe and continuous localised spontaneous throbbing sensation. The GDP had already accessed the tooth prior to referral. The clinical and radiographic examination was completed as per the protocol previously outlined. Tooth 43 was diagnosed with previously initiated therapy and asymptomatic apical periodontitis. RCT was performed as per the protocol previously outlined in August 2019. A periapical radiograph revealed a satisfactory root filling and the patient is scheduled for review.

Case 2

A 26-year-old male was referred by their GDP for management of two abscessed molars with open apices. At the time of initial assessment in May 2014, the patient was asymptomatic but reported a history of receding gums, a broken tooth and multiple abscesses over the past year. Medically, there was a confirmed diagnosis of XLH with bilateral deformities of both tibias and a previous fracture of the left tibia. The patient's medications included Calcitriol and K-Phos. The patient's dental history revealed that he was an irregular attender previously whilst at university and had frequently moved between dental practices due to relocation. The patient reported a family history of XLH.

Extra-oral examination was unremarkable. Intra-oral examination revealed generalised erosion (Figure 3a & 3b). A comprehensive endodontic examination performed according to the previously outlined protocol revealed unrestored teeth 24 and 46 with associated draining buccal sinuses (Figure 4a) and a fractured tooth 26 with a temporary dressing *in situ* (Figure 3a). All three teeth gave negative responses to cold testing. Radiographic examination, consisting of two periapical radiographs using standard settings (0.4 s exposure time, 70 kVp and 7 mA), was performed to assess the root canal morphology and the presence of apical pathosis (Figure 4b). Tooth 46 demonstrated a high mesial pulp horn, short roots with possible root resorption and open apices with associated periapical radiolucencies. A Cone-Beam Computed Tomography scan (CBCT) (Orthophos XG 3D, Dentsply Sirona, Charlotte, NC, USA) with standard settings (5.1 s exposure time, 85 kV and 7 mA) of tooth 46 was undertaken in June 2014 due to its unusual anatomy, especially the furcal wall of the mesial root, to guide endodontic treatment (Figure 5). The scan was reported by a consultant dental and maxillofacial radiologist using the software described above. The report indicated the presence of only two canals, considerable loss of buccal and interradicular bone and an appearance suggestive of external root resorption. The diagnoses were pulp necrosis and a chronic apical abscess of teeth 24 and 46 and asymptomatic apical periodontitis of tooth 26.

Verbal, informed and valid consent was obtained to perform RCT on teeth 24, 26 and 46, which was performed on tooth 24 in March 2015 as per the above protocol outlined in case 1. Tooth 26 required root filling with a combination of techniques that was completed in April 2015. The palatal canal of tooth 26 was filled with an MTA plug and gutta-percha backfill and the mesio-buccal and disto-buccal canals were filled with a warm vertical compaction technique. While treating tooth 24, a sinus was noted associated with tooth 22. At this point, sensibility testing of the full mouth with an electric pulp tester (Sybronendo, Orange, CA, USA) was performed which demonstrated no response from teeth 22, 25, 27, 32 and 31. This presented a diagnostic challenge, but with a lack of signs and symptoms from teeth 25, 27, 32 and 31, it was decided to keep these under close observation. The diagnosis for tooth 22 was confirmed as pulp necrosis and a chronic apical abscess and RCT was completed in May 2015. Tooth 46 was filled entirely with MTA in August 2015 due to its unusual anatomy and a post-operative periapical radiograph revealed a satisfactory root filling (Figure 4c).

Following completion of the initial treatment plan, the patient reattended in January 2016, May 2016, May 2017 and September 2018 with new complaints of pain and swelling. As a result, following appropriate investigations and diagnosis, RCT was performed on teeth 14, 21, 23, 32, 31 and 41. The pre-operative periapical radiograph of tooth 14 demonstrated a widened periodontal ligament with complex root canal anatomy and a suspected apical trifurcation (Figure 6a). Clinically, an apical bifurcation was detected and this was filled with a warm vertical compaction technique with a periapical radiograph demonstrating a satisfactory root filling (Figure 6b). The pre-operative periapical radiograph of teeth 32, 31 and 41 demonstrated high pulp horns close to the amelo-dentinal junction, apical radiolucencies and a GP cone tracing through a sinus tract to locate the source of infection (Figure 7a). Canal filling was performed with a warm vertical compaction technique and a periapical radiograph demonstrated satisfactory root fillings, albeit with some extrusion of sealer (Figure 7b).

On review in September 2018 and October 2019, all endodontically treated teeth were asymptomatic and clinically there were no signs of infection or periapical inflammation with no adverse consequences. Radiographically, there was evidence of normal periodontal ligament spaces around teeth 14, 21, 22, 23, 26, 32, 31, 41 and 46 (Figure 4d, 6c & 7c) and the outcome was considered favourable (European Society of Endodontology 2006). The apical radiolucency associated with tooth 24 had diminished in size but after review at 4 years 7 months, in the presence of a residual apical area, the outcome was considered unfavourable and endodontic retreatment is being considered (European Society of Endodontology 2006). The patient is now being kept on a 6-monthly recall.

Discussion

The two case reports demonstrate the development of ‘spontaneous’ pulpal necrosis and apical pathosis associated with intact teeth in two young male patients. Both patients had a confirmed diagnosis of XLH and a strong family history. They presented with a caries free dentition and no history of trauma. It is an unusual presentation to find pulpal and apical pathosis in intact teeth.

Yet, this presentation is consistent with other published case reports of patients with XLH (Murayama *et al*. 2000, Alexander *et al*. 2001, Pereira *et al*. 2004, Beltes & Zachou 2012, Lee *et al*. 2017, James & Roudsari 2019).

Patient 2 experienced more widespread dental disease and required RCT on 10 permanent teeth. Some reports suggest that males with XLH tend to present with more severe dental disease than females (Holm *et al.* 2001), while others propose no difference (Andersen *et al.* 2012). The majority of teeth affected were anterior teeth in the cases reported here. This is consistent with the findings from Andersen *et al.* (2012) in their cross-sectional study which reported a predominance for anterior teeth, most commonly incisors followed by canines. They also found only incisors and canines were affected in younger patients, consistent with patient 1, and the proportion of posterior teeth affected increased with age, consistent with patient 2. The more extensive disease experience of patient 2 may be explained by the 10-year age gap between the patients allowing more time for the development of cracks, tooth wear and subsequent bacterial penetration. This finding is again consistent with Andersen *et al.* (2012) who demonstrated the number of affected teeth rose significantly with age. Another possible explanation may be the extent of tooth wear which could potentiate the development of dental infection through cracks and thinning of the enamel; patient 2 presented with erosive tooth wear whereas patient 1 had no visible enamel defects. Nonetheless, Goodman *et al.* (1998) found that in abscessed permanent incisors where significant tooth wear was absent, enamel infractions were often present. Still, they recognised that in some cases there did not appear to be any logical explanation demonstrating the uncertainty in the pathogenesis. This is acknowledged by Hillmann & Geurtsen's (1996) histologic examination which demonstrated the invasion of microorganisms into enamel without any visible surface defects.

Diagnosis was found to be a significant challenge in these patients owing to the absence of observable dental disease. The spontaneous presentation of endodontic infection means it is impossible to predict future infection from clinical examination alone. Full mouth sensibility testing was performed on patient 2 with the aim of identifying teeth with pulpal necrosis, an approach also adopted by Alexander *et al*. (2001). Four intact teeth had demonstrated a negative response but with an absence of any symptoms or clinical and radiographic signs of infection. This is in contrast to Alexander *et al*. (2001) where periapical lesions were also visible radiographically on those teeth which demonstrated a negative response. In addition, the patient subsequently developed pulpal necrosis in teeth which had tested positive. Previous studies have found there to be a lack definitive correlation between the results of sensibility testing and the histological status of the pulp, adding to the diagnostic challenge (Seltzer *et al.* 1963, Dummer *et al.* 1980). Although more recent studies show a greater agreement, they still advocate an improved method of obtaining a reliable pulp diagnosis (Ricucci *et al*. 2014). It was acknowledged that no single diagnostic test should be relied upon and sensibility testing should only be used for diagnosis where other tests were concordant (Dachi *et al*. 1967). Consequently, it was decided to only treat symptomatic teeth or those demonstrating obvious clinical signs of infection to avoid unnecessary intervention.

Amongst both patients, the full array of unusual morphological features quoted across the literature within one case report were demonstrated. Enlarged pulp chambers and pulp horns seem to be the most commonly reported findings (Hillmann & Geurtsen 1996, Murayama *et al*. 2000, Alexander *et al*. 2001, Pereira *et al*. 2004, Souza *et al*. 2010, Lee *et al*. 2017). Enlarged pulp chambers were found in many of the teeth with necrotic pulps with close extension to the amelo-dentinal junction in teeth 32, 31 and 41 of patient 2. Patient 1 presented with a 'thistle shaped' pulp chamber on tooth 21 which has seldom been reported in these patients (Goodman *et al.* 1998). Enlarged root canals are also another feature that was identified, particularly apparent in tooth 21 of patient 1, similar to cases found by Pereira *et al.* (2004) and Lee *et al.* (2017). Open apices appear to be a less commonly reported in the literature (Pereira *et al*. 2004), but these were present in both teeth 21 and 43 in patient 1 and in teeth 21, 24, 26 and 46 for patient 2. The open apices may be attributable to the young presentation of patients where root formation is incomplete at the time of pulpal necrosis. Short roots and root resorption complete the range of features reported previously and this was identified to be the case in tooth 46 of patient 2 (Hillmann & Geurtsen 1996, Murayama *et al*. 2000). An apical trifurcation of tooth 14 of patient 2 was suspected and clinically an apical bifurcation was detected. Although it was appreciated a CBCT would have revealed the true anatomy, this would not have altered the management and therefore the exposure was not justified in this instance.

The unusual morphology adds to the complex management of these patients. To the best of our knowledge, one published case report demonstrates an apexification technique with calcium hydroxide, however, following treatment, the sinus tract persisted and surgery was subsequently required (Pereira *et al*. 2004). Despite the array of literature on MTA, no studies document its application in patients with XLH. The application of MTA in cases of wide canals and open apices has been shown to give favourable clinical outcomes (Mente *et al*. 2013, Torabinejad *et al.* 2017). In this case report the application of MTA in two patients is described and favourable outcomes have been demonstrated. A potential limitation of the case report is the lack of longer-term follow-up. It is plausible that post-treatment disease may occur in well disinfected and filled canals through bacterial invasion of defective dentine. Therefore, full coverage restorations could be considered for these patients to cover exposed dentine. Closer follow-up would also be recommended. Over a 4-year follow-up of tooth 24 in patient 2, an unfavourable outcome was demonstrated, and although retreatment is being considered, this poses many treatment challenges. Firstly, the use of an MTA plug makes removal difficult, risks thinning the dentine walls and extrusion of MTA debris. Furthermore, endodontic microsurgery has its limitations in this case due to the short root length.

It has been widely acknowledged that prevention of such presentations should be at the centre of care for patients with XLH. This should commence from a young age and patients should be encouraged to attend for regular dental examinations with their GDP (Souza *et al.* 2010, Opsahl Vital *et al*. 2012). Regular professional cleaning, topical fluoride applications and maintenance of good oral hygiene are considered essential (Seow 2003, Souza *et al.* 2010). Interventions to prevent bacterial penetration and preserve pulp vitality should be performed. Suggested interventions include the provision of fissure sealants and stainless-steel crowns on posterior teeth and composite coverings on anterior teeth (Seow 2003, Souza *et al.* 2010). It is recognised that early supplementation and good compliance with calcitriol and phosphate during the development of the permanent teeth can improve dentine mineralisation reducing the risk of developing dental abnormalities and spontaneous abscesses (Chaussain-Miller *et al.* 2007, Opsahl Vital *et al*. 2012, Beck-Nielsen *et al*. 2019).

Conclusion

Patients diagnosed with XLH may present with spontaneous pulpal necrosis and apical pathosis in intact teeth with no history of trauma, posing diagnostic challenges. The range of abnormal root canal morphology may present endodontic challenges which can be managed effectively with an appropriate root canal treatment protocol. Positive treatment outcomes have been demonstrated, although it is clear that further research into prevention protocols will be key to avoiding such presentations.

Conflict of interest

The authors have stated explicitly that there are no conflicts of interest in connection with this article.

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Figure Legends

Figure 1 PRICE 2020 flowchart

Figure 2 Periapical radiographs demonstrating RCT of tooth 21. (a) Pre-operative periapical radiograph demonstrating unusual coronal anatomy, a wide root canal space and apical pathosis. (b) Immediate post-operative periapical radiograph demonstrating a satisfactory root filling of an MTA apical plug, gutta-percha backfill and restored with bulk-fill and nanohybrid composite. (c) 8-month review periapical radiograph demonstrating complete resolution of the apical radiolucency and the presence of a normal periodontal ligament space.

Figure 3 Intra-oral colour photographs taken at initial assessment. (a) Upper occlusal demonstrating a fractured tooth 26 and generalised erosion. (b) Lower occlusal demonstrating generalised erosion.

Figure 4 Intra-oral colour photograph and periapical radiographs demonstrating the presentation and RCT of the 46. (a) Draining buccal sinus associated with the intact tooth 46. (b) Pre-operative periapical radiograph demonstrating a high mesial pulp horn, short roots, possible root resorption and open apices with associated periapical radiolucencies. (c) Immediate post-operative periapical radiograph demonstrating a satisfactory root filling with MTA. (d) 48-month review periapical radiograph demonstrating complete resolution of the apical radiolucencies and the presence of a normal periodontal ligament space.

Figure 5 CBCT taken to assess the unusual anatomy of tooth 46. (a) Axial section demonstrating unusual root anatomy associated with the mesial root of tooth 46, with narrowing of dentinal walls apically and evidence of external root resorption. (b) Sagittal section demonstrating loss of the interradicular bone and evidence of external root resorption. (c) Coronal section demonstrating unusual course of root canal in mesial root of tooth 46, along with possible external root resorption affecting mesial aspect of the root in coronal to middle third.

Figure 6 Periapical radiographs demonstrating RCT of tooth 14. (a) Pre-operative periapical radiograph demonstrating a widened periodontal ligament with a suspected apical trifurcation. (b) Immediate post-operative periapical radiograph demonstrating satisfactory canal filling of an apical bifurcation with a warm vertical compaction technique. Image cropped to allow comparison to pre-operative image. (c) 31-month review periapical radiograph demonstrating complete resolution of the apical radiolucency and the presence of a normal periodontal ligament space. Image cropped to allow comparison to pre-operative image.

Figure 7 Periapical radiographs demonstrating RCT of teeth 32, 31 and 41. (a) Pre-operative periapical radiograph demonstrating high pulp horns close to the amelo-dentinal junction, apical radiolucencies and a GP cone tracing through a sinus tract to locate the source of infection. (b) Immediate post-operative periapical radiograph demonstrating satisfactory root fillings using a warm vertical compaction technique with some extrusion of sealer. (c) 22-month review periapical radiograph demonstrating complete resolution of the apical radiolucencies and the presence of a normal periodontal ligament space.

PRICE 2020 Flowchart

2 male patients: **Patient 1** age 16; **Patient 2** age 26

PC: Recurrent infections

Verbal, informed and valid consent for investigation of complaint

MH: XLH

PDH: **Patient 1** recurrent infections in the primary dentition

Buccal swellings and draining buccal sinuses on intact teeth

Patient 1: Teeth 21, 43; **Patient 2:** Teeth 14, 21, 22, 23, 24, 26, 32, 31, 41, 46

Patient 1: IOPAs; **Patient 2:** IOPAs & CBCT

High pulp horns, short roots, open apices and periapical radiolucencies

No response to pulp sensibility testing (cold)

Diagnoses:

Patient 1: Pulp necrosis and chronic apical abscess: 21; Previously initiated therapy and asymptomatic apical periodontitis: 43

Patient 2: Pulp necrosis and symptomatic apical periodontitis: 14, 21, 23; Pulp necrosis and chronic apical abscess: 24, 46, 22, 41; Pulp necrosis and asymptomtic apical periodontitis: 26, 32, 31

Verbal, informed and valid consent for treatment

Intervention: NSRCT. MTA plug: **patient 1** teeth 21, 43; **patient 2** teeth 21, 23, 24, 26, 46

Follow-up: range 8m - 4 yrs+

Follow-up assessment: IOPA, percussion and palpation tests, presence or absence of sinus, mobility and periodontal probing depths

Treatment outcome: symptom-free, clinically no signs of infection or inflammation, radiographic evidence of resolution of periapical radiolucency, except patient 2 tooth 24 where radiolucency diminished in size after 4 years

Patient perspective: pain-free and could function normally

Conclusions: Patients with XLH may present with 'spontaneous' pulpal necrosis. The abnormal root canal morphology can be managed effectively with NSRCT

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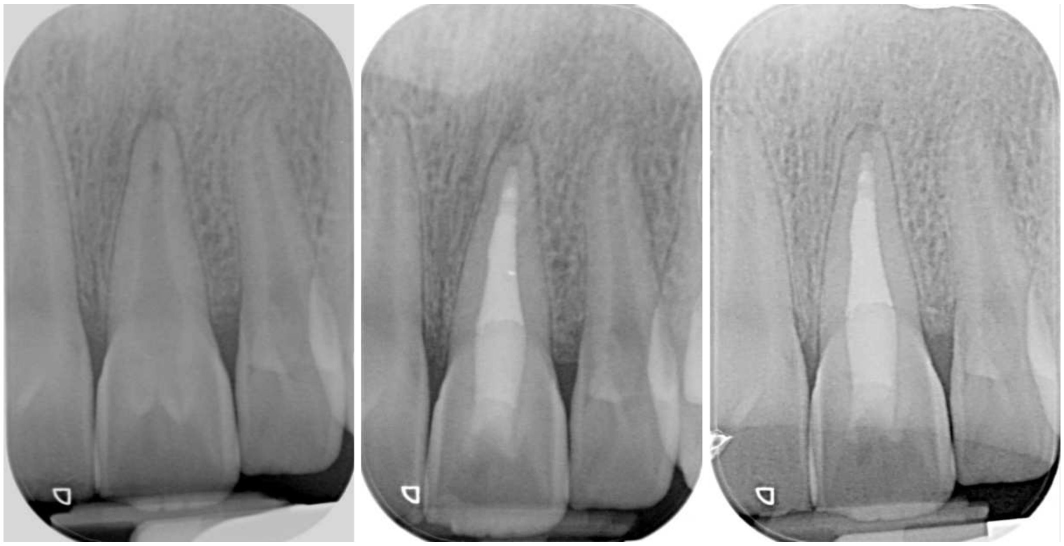
Author has started explidtly that there is no conflict of interest

***From: Nagendrababu V, Chong BS, McCabe P, Shah PK, Priya E, Jayaraman J, Pulikkotil SJ, Setzer FC, Sunde PT, Dummer PMH (2020) PRICE 2020 Guidelines for reporting case reports in Endodontics: A consensus-based development. *International Endodontic Journal* doi: 10.1111/iej.13285.**

For further details visit: http://pride-endodonticguidelines.org/price/



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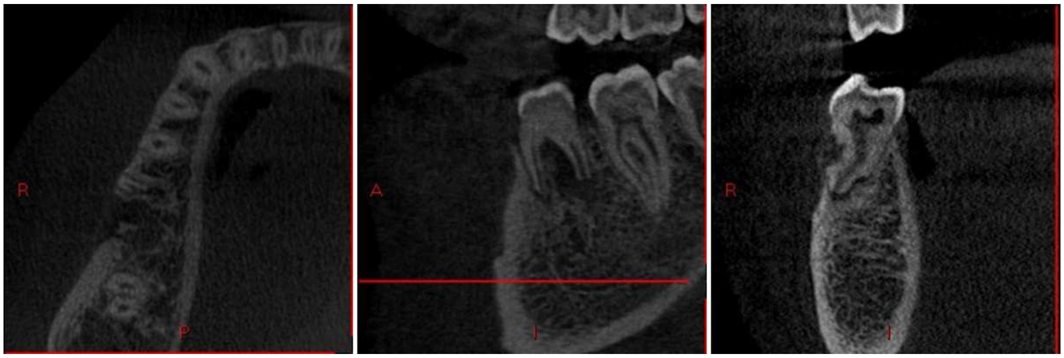

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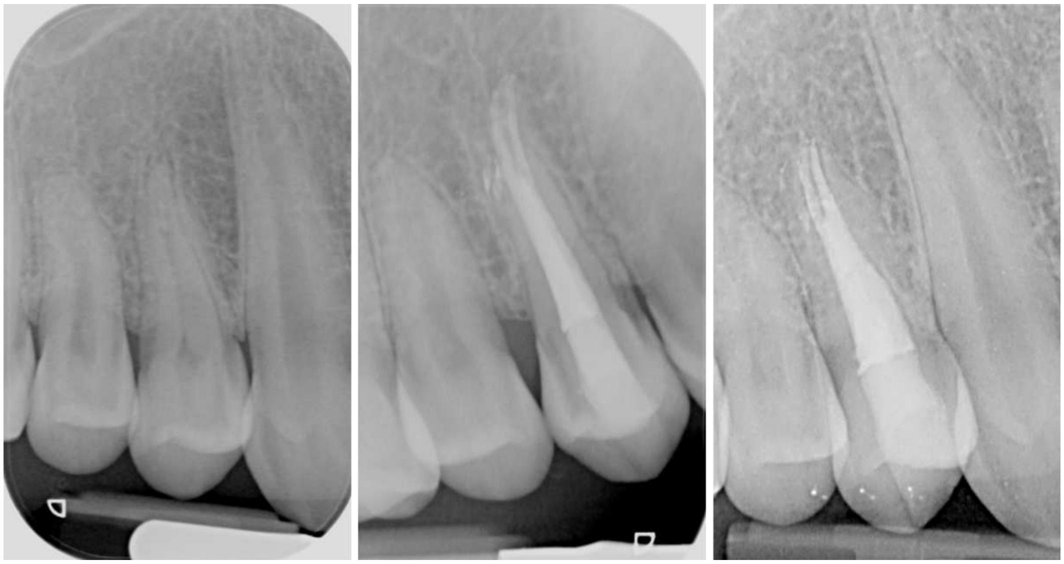

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