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Dental treatment of children with krabbe syndrome with tracheostomy - Clinical case report

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Abstract*—*People with special needs, have momentary or permanent limitations, one or more barriers, with physical, motor, cognitive, mental or sensory biological etiology. Dentistry for patients with special needs is the only specialty that works with all ages and is present in the three strands of dental care at home, outpatient and hospital allowed by the federal council. Krabbe Syndrome is an autosomal recessive neurodegenerative disease resulting from a deficiency of the enzyme galactosylceramidase, affecting the demyelination of the central and peripheral nervous system. In the literature, there are no reports on outpatient dental care for people with tracheostomy or Krabbe's disease. Given this, the objective of this study is to monitor and treat the oral health of a child with a rare syndrome with tracheostomy, who can have good oral health free of caries and periodontal disease. Thus, bringing the benefit of accessibility to outpatient dental care for children with tracheostomy, avoiding hospital interventions.

I. INTRODUCTION

According to the Brazilian Law for the Inclusion of Persons with Disabilities, in article 2, a person with a disability is considered a long-term impediment with etiology of a physical, mental, intellectual or sensory nature that they have, which, in interaction with one or more barriers, can obstruct their full and effective participation in society on equal terms with other people. Based on this, the demographic census (2019) released by the Brazilian Institute for Static Geography (IBGE), indicates that approximately 45 million people have some type disability, that is, 25% of the Brazilian population 1 .

The Ministry of Health and the Federal Council of Dentistry recognize that dentistry for patients with special needs individuals with simple or complex changes, momentary or permanent, biological, physical, mental, social and/or behavioral etiology that require a special, multidisciplinary approach and protocol specific[1,2].

Krabbe syndrome is an autosomal recessive neurodegenerative disease characterized by a deficiency of the enzyme galactocerebrosidase3,4. Described in 1916 by neurologist KnudHaroldsenKrabbe, he observed neuropathological characteristics in five children with diffuse sclerosis in the brain [3].

Rare disease with an estimated incidence of 1:100,000 live births3. It can be classified into two clinical forms: early childhood and late[3,4]. Main symptoms are: excessive crying, irritability, stiffness, seizures, motor delay, dysphagia, seizures, loss of vision[3,4]. With poor prognosis and rapid evolution, the life success rate is almost impossible[3,4].

The dysphagia of the disease leads to the need for tracheostomy, a surgical procedure that aims to unblock the airways[5,6]. It can be performed at any age, it is classified as temporary or permanent, depending on the patient's disability[6]. In most cases, feeding is compromised and there may even be the need for an enteral tube. Regardless of the route and nutrition, oral health and cleaning of the tracheal tube must be redoubled [23,13].

In the oral cavity, there are over 700 species of bacteria that make up the oral microbiota that act as beneficial and harmful pathogens [7]. In both situations, pH, presence of nutrients, salivary flow and retention factors that facilitate the maturation of the biofilm with the imbalance of a of these four pillars, the perfect opportunity for oral diseases can occur, they can lead to death [7].

Patients with special needs with motor difficulties and/or more time-consuming intellectual processing have poor or non-existent oral hygiene and do not have the necessary dexterity to use dental floss and brushing [8].

Given this context, the objective of this work through clinical report, is to show the preventive dental treatment of a person with Krabee syndrome with tracheostomy, performed in an outpatient setting, paying attention to the techniques of behavioral management, preventive care and treatments. Thus cooperating for a quality perspective of life for these individuals.

II. CASE REPORT

Patient SRO, leucoderma, female, 05 years old, tracheostomized, attended the dental clinic for Patients with Special Needs at the InstitutoBrasileiro do Norte (IBEN-AM), accompanied by a mother for dental treatment with the main complaint of gingival bleeding, bad breath and sporadic teeth grinding.

In the first consultation, an oral evaluation, a treatment plan, a clinical case study and exposure of biological images of the quarterly evolution of oral health for scientific purposes were requested and authorized by means of an informed consent form.

During the anamnesis, the mother reported that the patient is the first case of a person with a disability in the family, unplanned pregnancy, uneventful prenatal period that could present risks for the baby. In the postnatal period, around the age of two years, the child suffered trauma and, twenty-four hours after the event, developed a fever of forty degrees Celsius, loss of movement in the upper and lower limbs and regression in oral communication. The mother informs that the patient does not have systemic changes, she has neurological monitoring in the public health system every six months, she does not use continuous medication and does not have allergies. Their food is provided twice a day by nasogastric tube and orally

When asked about oral health care, the mother reported that she tried to brush at least once a day, never attempted to use dental floss and fluoride toothpaste. She breastfed exclusively until she was six months old and then introduced a bottle until she was two years old with a dairy compound.

On extraoral examination of the patient, there is a constant purulent secretion in his tracheal tubule, total motor dependence, involuntary, non-verbal spasms. (Figure 1 and 3).

On intraoral examination, open bite class II, mixed arch dental wear caused by bruxism, periodontal disease with presence of calculi and presence of active carious lesion.(Figure 2)

Defined treatment plan: prophylaxis, tartarectomy and fluoride application.

The organization of the bench and ergonomics during the service were adjusted to the patient's comfort. (Figure 4 and Figure 6)

Respecting and adapting the handicap of the disabled person at the beginning of the procedure, asepsis of the oral cavity was performed, using 0.12% chlorhexidine digluconate brand ORAL-B soaked in sterile gauze with the aid of a wooden tongue depressor made with a popsicle stick adhesive tape. (Figure 5)

All consultations were carried out in the patient's wheelchair, sometimes with the need for protective stabilization. Guidance to the mother and demonstration of the brushing technique. (Figure 6)

Preventive control appointments followed the sequence. Placement of a children's lip retractor brand indusbelo.

The prophylaxis was carried out with ALLplan prophylactic paste with strawberry flavor and sswhite pumice stone. Periodontal probing with the aid of the millimeter probe WHO, for subgingival and supragingival root planing, the instrumental gracey curettes 5-6,7-8,1-12,13-14, mini five curettes 5-6,7-8 were used ,11-12,13-14 and ultrasonic scallermicrodont device, sterile gauze, relative isolation for application of strawberry fluoride. (Figure 3)

In the first visit, caries was observed in the initial active process in element 53, with the application of fluoride varnish. The importance of oral care at home was

reinforced to family members for the regression of this caries. (Figure 8)

In the second visit, eruption of permanent teeth 11 and 21 is noticed. Patient without presence of carious lesion. With accumulation of dental calculus predominant in the posterior region. (Figure 9 and 10)

In this preventive return, element 52 was extracted naturally without anesthetic, as it was already in the exfoliative phase. On that day, the mother was explained how she should remove the other little teeth that were in the exfoliative phase. (Figure 11 and 12)

Following the treatment plan, this was the post pandemic consultation, with the birth of tooth 22, gum hyperemia and no oral disease. Current brushing happens three times a day, flossing with a cable was an efficient alternative for introducing this habit. (Figure 12 and 13)

The total treatment time was nine months of treatment without any pain or discomfort, with caries or periodontal disease being controlled, but with the presence of idiopathic gingival hyperplasia. (Figure 14)

Even with the uncontrollable spasms and the outpatient setting, it was possible to carry out the proposed treatment plan, with a favorable prognosis. Creating a daily care routine with the help of wood style opener and 0.12% chlorhexidine, small round head soft toothbrush and fluoridated toothpastes. (Figure 1)

During the consultations, difficulties were faced, such as the excess of dental calculi that were supported with the aid of a diaper that the patient uses to control the excretion of the tracheal tubule so that there was no obstruction of the tracheal tube. Especially the communication disorder that did not allow the patient to interact with the professional and the dental environment.

III. FIGURES

Fig. 1: Explanation of stabilization for oral health care.
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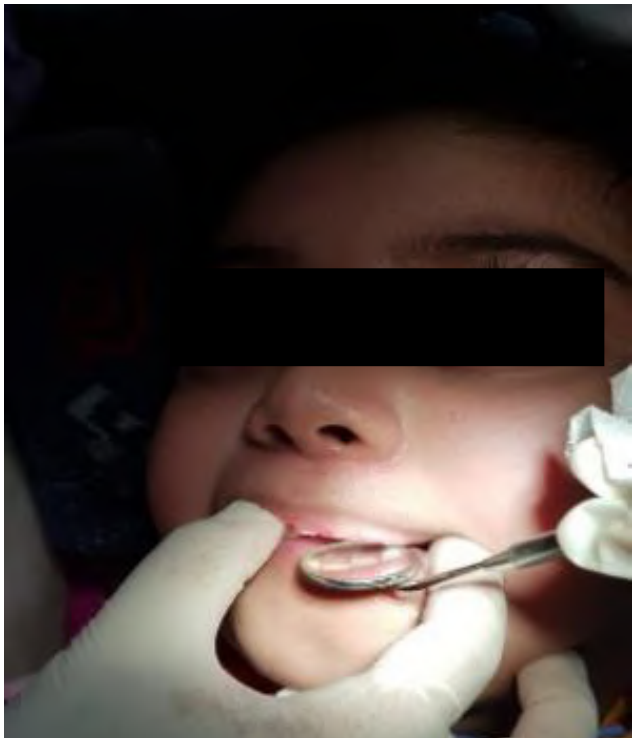

Fig. 2: Initial clinical examination.
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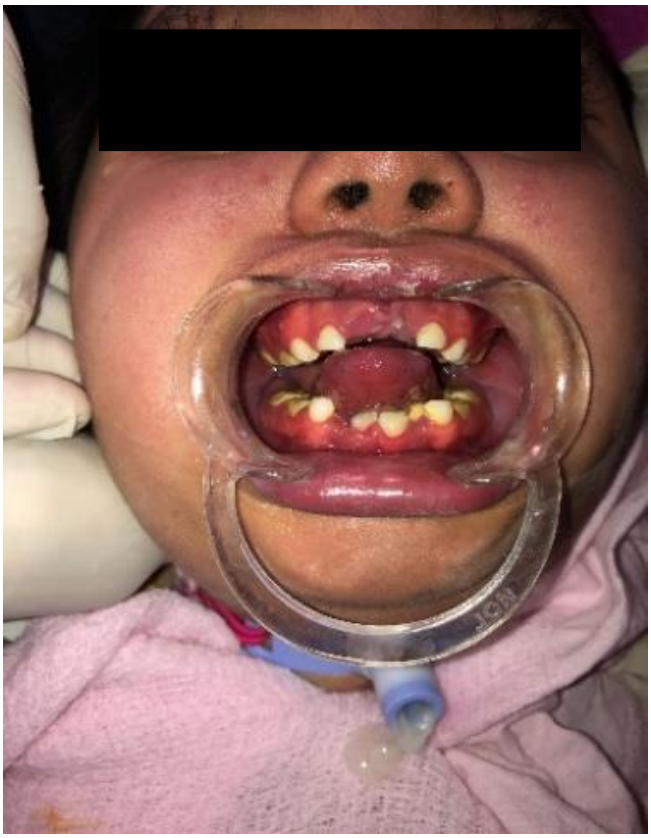

Fig. 3: intraoral analysis and secretion control.
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Fig. 4: Organization of preventive care workbench.
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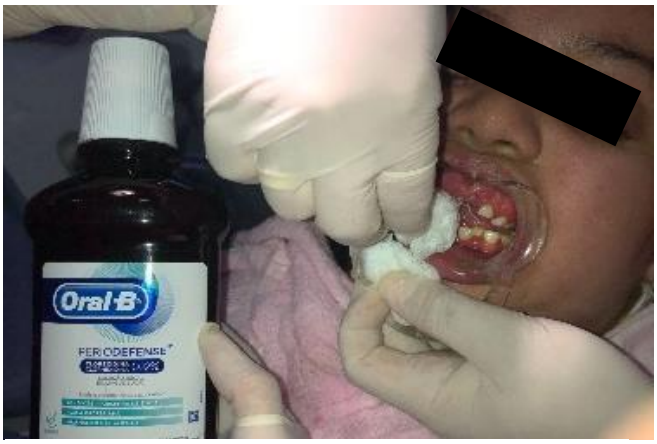

Fig. 5: Intraoral asepsis
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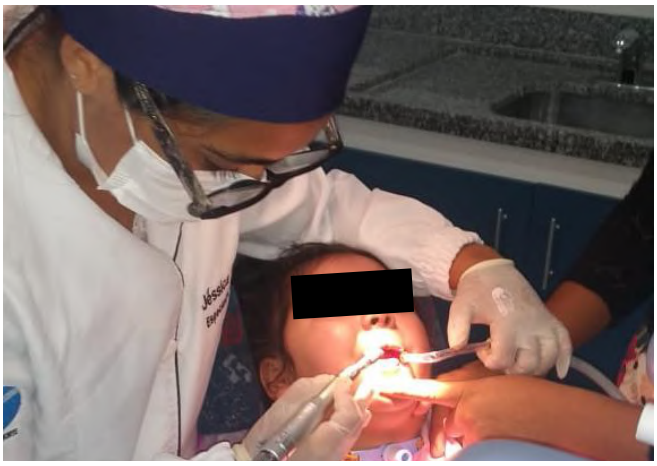

Fig. 4: Organization of preventive care workbench.
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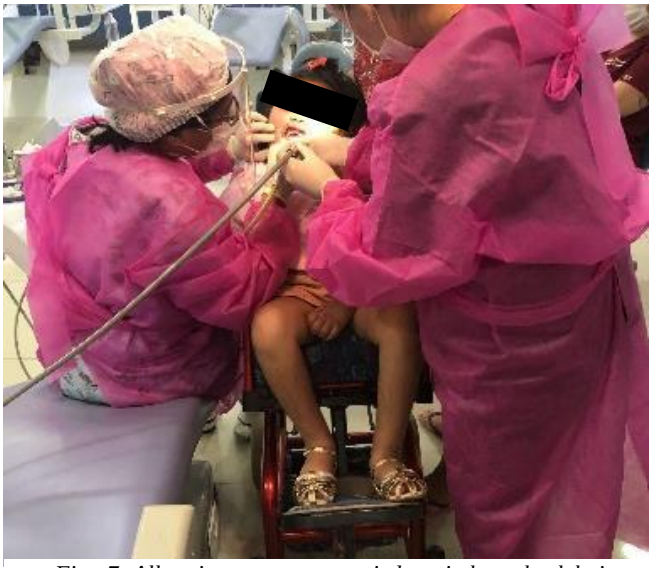

Fig. 7: All patient care was carried out in her wheelchair
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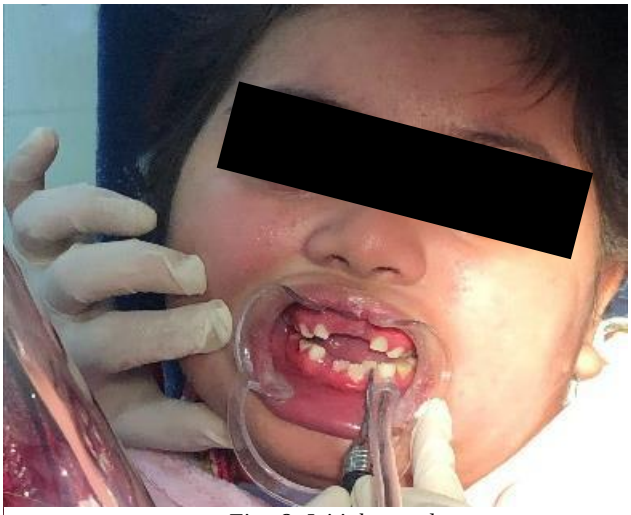

Fig. 8: Initial record.
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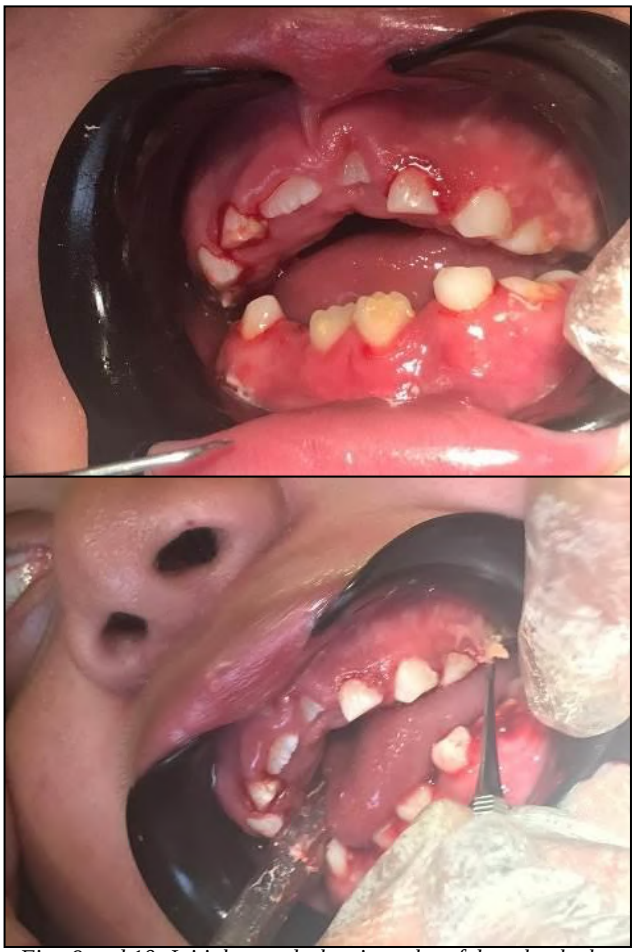

Fig. 9 and 10: Initial record, showing a lot of dental calculus being removed
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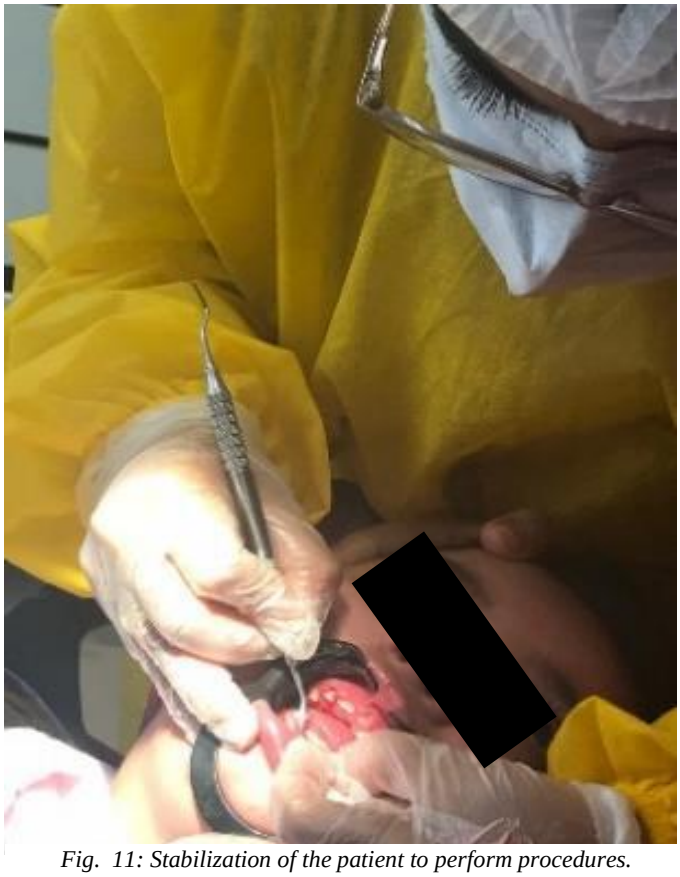

Fig. 11: Stabilization of the patient to perform procedures.
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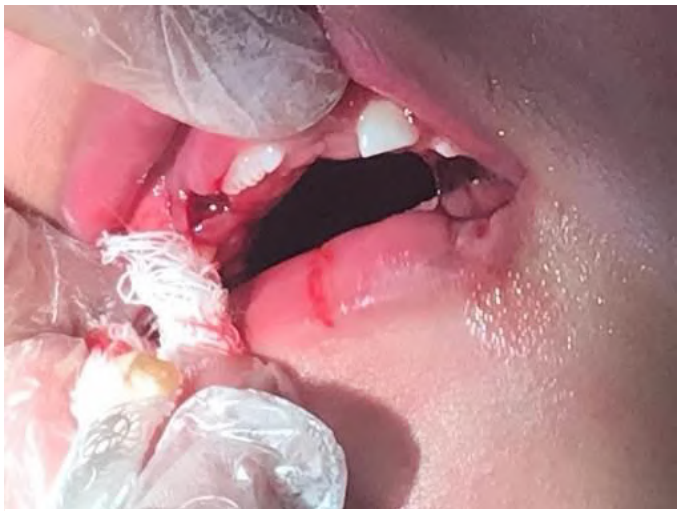

Fig. 12: Deciduous dental element extraction (52).
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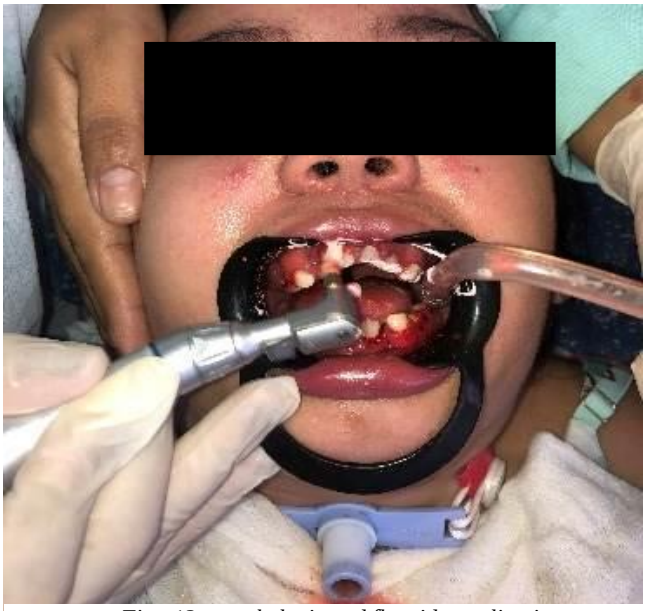

Fig. 13: prophylaxis and fluoride application.
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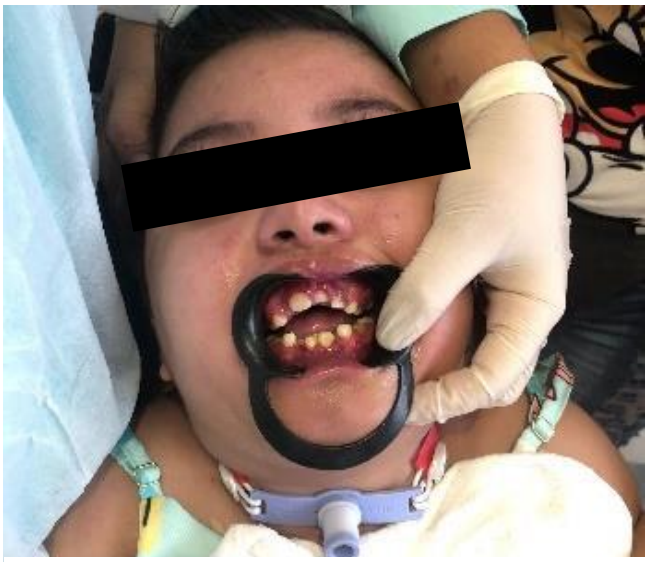

Fig. 14: Prophylaxis and fluoride application.
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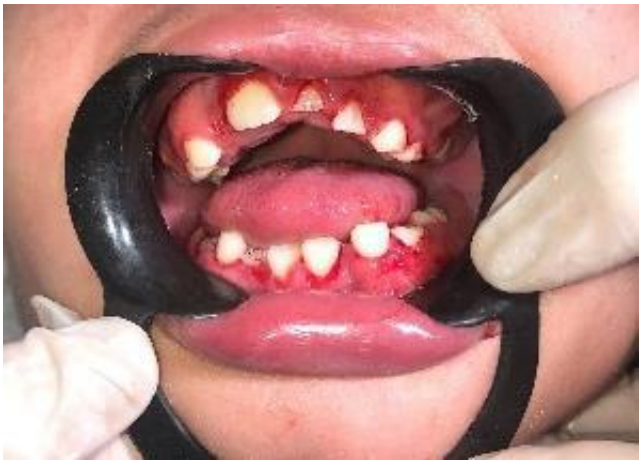

Fig. 15: Final aspect.
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IV. DISCUSSION

Krabbe's Syndrome is a degenerative leukodystrophy[4]. It has a difficult prognosis and rapid neurological degeneration, which causes muscle spasm, visual impairment, irritability, uncontrollable crying, respiratory difficulty. The average lifespan of these people is twenty-four months[3,4].

In the analyzed articles, it can be seen that the need for tracheostomy is fundamental for the quality of life of these individuals who have respiratory difficulties[25,5,16]. However, in the literature, in all case reports, patients became residents of hospitals and the nursing team was responsible for oral hygiene care in the hospital environment, differently from the report.

The first dental appointment in a human being's life should happen around 6 months of age, but children with special needs are rarely taken to the dentist early, mainly due to lack of encouragement from the medical team 10.9,

It is important that dental surgeons master the behavioral management technique to avoid skipping necessary oral health education steps. Refer to general anesthesia only when all methods are ineffective[10,11,7,9]. People with Down's Syndrome, for example, when undergoing general anesthesia may present some complications because they have a short neck and macroglossia, making trachial incubation difficult[11].

It emphasizes that people with Krabbe's disease who are discharged from hospital must be monitored by a multidisciplinary team with physiotherapists, nurses and medical staff, to maintain respiratory functionality[3]. Perez et al (2013), emphasizes the importance of oral health care for patients with neurodegenerative diseases with tracheostomy, in the prevention and control of infectious foci of oral origin that favor the risk of respiratory complications, which can lead people with disabilities to death.

Nogueira JWS, Jesus CAC (2017) and BASSAN LT et (2018), both demonstrated in a literature review format that the oral hygiene care protocol for patients who use mechanical ventilation in pediatric and adult intensive care units use brushing with toothpaste to control biofilm when they present teeth and use of 0.12% chlorhexidine gluconate antiseptic antimicrobial to avoid ventilator pneumonia and possible systemic infections.

Scientifically proven that people with special needs are more prone to oral diseases such as tooth decay and periodontal disease. Such risk factors are aggravated in the disabled by the use of baby bottles, cariogenic diet and lack of office visits[15]. Therefore, it is up to the dental surgeon to show, perform and monitor guardians and/or

caregivers in maintaining the oral health of their dependents[15, 16.17].

V. CONCLUSION

The present work showed the efficiency and possibility of preventive control in the outpatient setting of people with Krabs Syndrome with tracheostomy, avoiding hospital interventions.

According to the studies carried out and the follow-up of the case report for nine months, there was a gap in the scientific literature on the care of people with rare diseases in an outpatient setting.

REFERENCES

[1] Lei Brasileira de Inclusão da Pessoa com Deficiência(Estatuto de Pessoa com Deficiência), Lei 13.146. 2013

[2] Campos CCC, et al. Manual prático para o atendimentoodontológico de pacientes com necessidadesespeciais. Ministério da SaúdeUniversidade Federal de Góias. Triagem:2°edição 2009.

[3] Lima K M et al. AbordagemmultiprofissionalnaDoença de Krabbe: relato de caso. Rev Neurocienc2021;29:1-9

[4] Sano T S .Doença de Krabbe: a importância do diagnósticoprecoce para seuprognóstico. Krabbe disease: the importance of early diagnosis to prognosis. Einstein, 2012;10(2):233-5.

[5] Cruz F S et al. Traqueostomia - Condutas e Técnica. Rev. Cir. Traumatol. Buco-Maxilo-Fac., Camaragibe v.20, n.2, p. 40-44, abr./jun. 2020 Brazilian Journal of Oral and Maxillofacial Surgery – BrJOMS.

[6] Teixeira, L A S, Castro MCS. Pacientes com traqueostomia: conhecimentos, atitudes e práticas das equipes do serviço de atençãodomiciliar. RevistaSustinere, Rio de Janeiro, v7,n2,p324-361, jul-dez,2019.

[7] Herculano, A B S. et al. Qualidade de saúdebucal e pneumonia associada à ventilaçãomecânica. Arch Health Invest 6(7) 2017

[8] Nunes R. et al. Prevalência de alteraçõesbucaisempessoas com deficiênciaclínica da universidade do extremosulcatarinense. Oral changes prevalence in persons with disability at the clinic of universidade do extremosulcatarinense. Odontol. Univ. Cid. São Paulo 2017; 29(2): 118-28,mai-ago ISSN 1983.

[9] Cerqueira, RB, Vicentini A N, Botelho MPJ. Utilização de técnicas de manejoinfantil para o atendimentoambulatorial de pacientes com hidrocefalia. 2012 Anais Eletrônico VI MostraInterna de Trabalhos de IniciaçãoCientífica ISBN 978-85-8084-413-9.

[10] Perez, MOD et al, Abordagemodontológicaemumapaciente com doençaTay-Sachs: relato de caso. Rev. Bras. Pesq. Saúde, Vitória, 15(2): 112-115, abr-jun, 2013

[11] Ussui A, et al. Característicasbucais e manejo com comportamental de pacientes com Síndrome de Down. e-Acadêmica, v. 1, n. 3, e15, 2020 (CC BY 4.0).

[12] Gonçalves, GC. Qualidade de vida da pessoa com traqueostomianaárea do grande Porto. Dissertação de Mestrado 2012. Site https://comum.rcaap.pt/handle/10400.26/9367.

[13] Nogueira JWS, Jesus CAC. Higienebucal no pacienteinternadoemunidade de terapiaintensiva: revisãointegrativa. Rev. Eletr. Enf. [Internet]. 2017 [acessoem: 28/08/2021];19:a46. Disponívelem: http://doi.org/10.5216/ree.v19.4148

[14] BassanLT,Peres MPS, Franco JB. Oral care in prevention of ventilator-associated pneumonia in neonatal and pediatric intensive care unit: protocol proposal. Rev. Bras. Odontol. 2018;75:e1183

[15] Resende VLS, Castilho LS, Viegas CMS, Soares. Risco para a CárieemDentesDecíduos de Portadores de NecessidadesEspeciais Risk Factors for Caries in Temporary Teeth in Disabled Children MA 1519-0501. 2007.

[16] Miranda AF, Sousa LB, Piauilino AIF. Intervençãoodontológicaempaciente com síndromerara – Doença de Huntington Dental Intervention To The Patient With Rare Syndrome. R CO. 2017;1(2):6-11.

[17] Chirano, CSB, Resende GB, Ribeiro EOA, Soares KS. Síndrome de Rett: abordagemodontológicaChirano, CSB. et al. RvAcBO, 2019; 8(2):15-19.

[18] PREVITALI EF, Ferreira MCD, Santos MTBR. Perfil dos Pacientes com NecessidadesEspeciaisAtendidosemumaInstituição de Ensino Superior Privada Profile of Special Needs Patients Assisted at a Private Higher Education Institution. Pesq Bras OdontopedClinIntegr, João Pessoa, 12(1):77-82, jan./mar., 2012 DOI: 10.4034/PBOCI.2012.121.12