

Comparative Analysis of the Muscular Structures of the Neck in Patients with Pierre Robin Sequence and Patients with Normal Mandibular Growth

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Abstract

Patients with Pierre Robin sequence have microretrognathia and studies prove that they never achieve normal mandibular development. These observations lean towards the idea that the suprahyoid muscles are shorter and play a crucial role in mandibular growth. Even though patients are treated with different surgical modalities, there is a tendency towards failed mandibular growth. Because of this, it's necessary to study the dimensions of the suprahyoid muscles that directly interact with the mandible. Besides, there are no quantitative reports of the degree of affectation of this muscle group. Using tomography images, the suprahyoid muscles in Pierre Robin patients were measured and compared to similar age patients without the anomaly. Results show that the most affected muscles are the geniohyoid and the anterior belly of the digastric muscle. This explains the natural tendency towards micrognathia and microgenia observed in these patients during development. The affectation of the mylohyoid explains the light concavity observed in the mandible body of Pierre Robin patients. This study proves the important role of the suprahyoid muscle group that, through their short length and contraction, secondarily affects mandibular growth.

Keywords: Pierre Robin Sequence, suprahyoid muscles, anterior belly of digastric muscle, mylohyoid, geniohyoid.

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1 Introduction

1.1 Definition

Pierre Robin sequence, also known as Pierre Robin malformation or Pierre Robin syndrome, is a congenital condition of facial abnormalities in humans characterized by a triad: an unusually small jaw (micrognathia), posterior displacement of the tongue (glossoptosis), and cleft palate. These conditions result in obstruction of the upper airway, from mild to severe, and the malformation is often accompanied by abnormalities in other anatomical regions. [1,2] Because the defect is not caused by a defect in a single gene, it is not a genetic syndrome, but a sequence: a chain of certain developmental malformations, one that leads to another and corresponds to a type of craniofacial syndromes classified as syndromes of the first brachial arch. [3]

1.2 History

The first case was described in 1822 by St. Hilaire. However, Robin gets credit for giving importance to this condition and noting the serious dangers that this entails.

Pierre Robin was a dentist born in France in 1867. He published in 1923 the first of 17 articles to glossoptosis. In 1929, he published a monograph on the subject. Robin described the feeding problems typically exhibited by these children and their difficulties in gaining weight. He also mentioned that glossoptosis could be the cause of cyanosis and pulmonary infection. In severe cases, he mentioned that death was inevitable and wrote, "I have never seen a child live more than 16 to 18 months presenting with hypoplasia with the mandible retracted over 1 cm behind the maxilla." [4,5]

1.3 Clinical Manifestations

In contemporary literature the clinical manifestations reported for this syndrome are micrognathia 91%, 70-85% glossoptosis, cleft palate (inverted U-shaped) 25%, 10-15% macroglossia and ankyloglossia, unusual nasal deformities, 10-30% ocular abnormalities. They may present with the following cardiovascular findings: innocent murmur, pulmonary stenosis, patent foramen ovale, atrial septal defect and primary pulmonary hypertension. 70 to 80% have abnormalities of the musculoskeletal system as syndactyly, dysplastic phalanges, polydactyly, clinodactyly, joint hypermobility and oligodactyly in upper limbs. In lower limbs: foot abnormalities, femoral malformations, anomalies hip, knee and tibia. You may also have spinal deformities such as scoliosis or kyphosis. [1,3,6,7,8].

1.4 Etiology

The specific cause of Pierre Robin sequence is unknown. The lower jaw develops slowly in the first months of life before birth, but growth accelerates in the first year. There are three theories to explain the pathogenesis :

Mechanical Theory : The most accepted. At some point in fetal bone formation, the tip of the jaw gets stuck at the sternoclavicular joint , restricting mandibular bone growth. It is believed that at 12 to 14 weeks of gestation, when the fetus begins to move, the

movement of the head causes the jaw to be released and from this moment it will grow again.

The mandibular hypoplasia (7th and 11th week of gestation) leads to a high tongue in the oral cavity, which prevents the tongue takes a natural position in utero on the floor of the mouth. This prevents abnormal palatal position valves are closed, leading to a cleft . This explains the classic inverted U cleft palate and absence of cleft lip. [9] Oligohydramnios can play a role in the deformation of the mandible. Low levels of amniotic fluid do not support the head in the uterus and this flexing allows the chin to move down against the chest, preventing normal mandibular growth. [10]

Neurological Maturation Theory: Delayed neurological maturation evidenced by electromyography of the tongue, palate and pharyngeal pillars, as a delay of the hypoglossal nerve conduction. Spontaneous correction in most cases, supports this theory. It has been reported that the commitment of the genioglossus muscle aggravates the obstruction.

Rhombencephalic Disneurological Theory: Regulatory Organization of motor and hindbrain relates to a larger problem of ontogenesis . [11]

Recently, a genetic cause of the sequence has been suggested, leading to a dysregulation in SOX9 and KCNJ2 genes. [12]

1.5 Associated Conditions

Pierre Robin sequence presents specific syndromes associated with 80% of cases. Commonly associated syndromes include Stickler syndrome (34%), 22q11 deletion (15%), fetal alcohol syndrome (10%), Nager, Treacher Collins and bilateral hemifacial microsomia. Children with Stickler syndrome have the same findings as the Pierre Robin sequence; also have flattened facies, myopia, skeletal changes as arachnodactyly, and mild to moderate arthritis. [2,13]

1.6 Incidence

The reported incidence varies from 1:8500 to 1:30000 live births. The gender distribution is 1:1. [14,15]

1.7 Diagnosis

Pierre Robin sequence may be suspected as early as 13 weeks of gestational age by ultrasonography and confirmed in the newborn by the manifestations already mentioned.

The newborn has an increased respiratory effort characterized by substernal, suprasternal and intercostal retraction. Also observed is growth failure, sometimes requiring enteral supplements, which can be provided by a catheter or gastrostomy tube. [7]The diagnosis of airway obstruction is established by physical examination and measurement protocols of blood oxygen with the child in a supine position. When children desaturate, arterial blood gases assess the degree of hypoxia and hypercapnia [8] The obvious changes include CO₂ retention with decreasing pH and compensatory elevation of seric HCO₃. [16] Polysomnography is used to differentiate between the central and obstructive apnea. Central apnea is considered if there is no associated muscular effort; and obstructive if muscular motion is detected in the absence of airflow. Infants with airway obstruction usually manifest at birth if this is a problem. Most of these children will outgrow the

problem at 3 months of age. [17] Any patient with Pierre Robin sequence should undergo genetic and ophthalmologic evaluation followed by regular eye examinations to limit ocular complications associated with Stickler syndrome . [2]

1.8 Complications

The most important medical problems are difficulty in breathing and eating. Infants who can not eat enough calories require dietary supplements. In these patients there are also risks of aspiration. Another adverse effect of chronic airway obstruction is hypoxia, hypercapnia and elevated pulmonary vascular resistance, which can lead to right heart failure. Associated mortality varies from 5 to 30% by Pierre Robin sequence in nonsyndromic patients mortality is 5.9%, and syndromic patients is 22.8% [16,17]

1.9 Management of Pierre Robin Sequence

Treatment goals focus on continued breathing, nutrition and optimize growth despite the willingness of breathing difficulties.

1.9.1 Conservative management.

The goal is to maintain a permeable airway and ensure adequate food intake. The supine position exacerbates airway obstruction and improves with prone position. An oxygen saturation monitor is used. If oxygen saturation is between 80 and 90 %, consider supplementary oxygen support. If the saturation is less than 80 %, consider intubation. If it is chronic, consider performing a tracheostomy. [8,17]

Mild cases can be treated effectively with correct positioning. There is evidence that cervical extension, plays a role in improving the airway. This position is maintained for 1-6 months to allow mandibular neuromuscular adaptation and growth. A nasotracheal tube can be placed as a temporary solution. [7] In moderate or severe cases, patients will require nasopharyngeal cannulation or placement of a nasopharyngeal tube to correct airway obstruction at the base of the tongue. Endotracheal intubation or tracheostomy may be required. In some centers lingual labial adhesion is done to bring the tongue forward. Mandibular distraction is effective in mobilizing the jaw forward to overcome the obstruction of the upper airway caused by the posterior position of the tongue. [18,19] The potential causes of airway obstruction include: structural pharyngeal abnormalities, tracheomalacia, acute angulation of the braincase, tongue abnormalities, subglottic abnormalities and hypotonia.

Furthermore, the placement of a nasogastric tube for food should be evaluated after 7 to 14 days, with serial weight measurements. Surgical treatment is indicated in patients who do not improve or present inability to control the movements of the tongue for 7 days or patients who can not be extubated in 3 days. [8]

The team involved in the education and management of the family or dependents of the child includes pediatricians, otolaryngologists, plastic surgeons, odonto-pediatricians, orthodontists, nurses, speech therapists, and social workers. This group ensures that each patient and family receives the most comprehensive plan using all available resources from birth to adolescence. [15]

1.9.2 Surgical Management

Treatment is guided by the severity of airway obstruction followed by the degree of difficulty in feeding. Children with pronounced micrognathia may have severe breathing difficulty or lack of weight gain. Surgical intervention is necessary in these cases. Various methods have been described, but the tracheostomy is the most used technique. Other surgical procedures, such as subperiosteal release of the floor of the mouth, and various types of glossopexy as Routledge technique or other forms of tongue - lip adhesion. The glossopexy must be released before an advanced dentition (9-12 months old) is developed. [20,21]

The gradual lengthening of the mandible by distraction osteogenesis is a technique that increases the size of the jaw at the expense of local bone. It is used in newborns with severe hypoplasia of the jaw that causes obstructive apnea. In adults the advance is 1 mm per day, however newborns can move from 2 to 3 mm per day since the bone heals quickly at this age. This also allows a gradual elongation of the soft tissues, muscles, vessels, nerves, skin, leaving the base of the tongue in a forward position and increases the dimensions of the retropharyngeal airway. It avoids performing a tracheotomy or maintaining prolonged intubation and significantly improves the quality of life. [18-22] Ow and Cheung reported in a meta-analysis that the most common cause of bilateral mandibular distraction is Pierre Robin sequence with 24.1% of cases. [23] New materials and techniques have improved elongation results. You can repeat the procedure if you need more bone elongation. The long term monitoring has shown that early bone distraction causes no sequelae nor interferes with the development of the teeth and/or jaw growth. It has become the procedure of choice in some centers.

Tympanostomy tubes are usually inserted when the palatoplasty therapy is performed to correct hearing loss by conduction and to prevent complications of the middle ear. [8]

Surgical procedures to repair the cleft palate (hard and soft) can be performed in one time. Most surgeons agree that the closure of the palate (palatoplasty) should be delayed until 15 to 18 months of age, even in the absence of ventilatory problems. If you previously performed a tracheostomy, palatoplasty can be performed safely and in any patient with cleft palate, between 10 and 12 months. [24] A direct relationship between the incidence of early postnatal obstructive postoperative complications and . Different early postnatal airway procedures have associated morbidity and mortality, so a thorough evaluation must be performed before choosing the appropriate technique. Tracheostomy for prolonged periods has been associated with significant morbidity such as tracheomalacia, chronic bronchitis, chronic lung disease and even sudden death (up to 64% complications in children under 1 year), all accompanied by very prolonged hospitalization .

The lingual adhesion should only be used as an interim procedure since it causes eating and speech disorders, as well as impaired tooth development in the affected area. The lingual traction using a Kirschner nail frequently produces lingual tears. Subperiosteal release of the mouth's floor musculature is not very effective. Patients with insufficient growth, often associated with malnutrition, increased respiratory morbidity and repeated hospitalizations, are candidates for a polysomnography study seeking pathological paths that could benefit from early treatment before debuting with more severe complications.

The criteria for surgery in these patients: breathing > 60/min, FIO₂ >60 %, PaO₂ <65 mmHg, PaCO₂ > 60 mmHg, weight gain of < 100 g / week, SaO₂ < 70 %.

1.10 Prognosis

Children affected by Pierre Robin sequence usually develop to reach full size. [25] Internationally, however, they have been found below the average size, which makes you wonder if the incomplete development is due to chronic hypoxia related to obstruction of the upper airway as well as lack of nutrition due to difficulties in early feeding and the development of an oral aversion. However, the overall prognosis is pretty good once the initial respiratory and feeding difficulties are overcome. Most babies with Pierre Robin sequence grow to lead a normal, healthy adult life.

Pruzansky and Richmond have shown that normal jaw growth can be achieved in a normal person and result in a normal profile at 4-6 years of age when the cause of micrognathia is deformational rather than a malformation. [26] Hanson and Smith found specific syndromes in 25% of their patients; multiple abnormalities without specific syndromes in 35%; and the remaining 40% have isolated Pierre Robin sequence.

1.11 Anatomy of the Neck's Anterior Region

The hyoid bone is an odd, symmetrical bone, transversely extended in the anterior neck. In the normal position of the head, its exact site corresponds to the sine of the angle between the inferior side of the face and the neck's anterior plane. It is positioned almost parallel to the lower edge of the mandible. Convex from the front and concave from the back, "U" shaped, so its name is derived from the Greek word *hyoeides* meaning "shaped like the letter upsilon" (υ). It consists of a body and four lateral extensions, two on each side, called cornu. Thirteen muscles attach to the hyoid. [27]

The neck has many important muscles whose primary role is to move the head, the cervical spine and the hyoid bone. These muscles are found symmetrically left and right of the cervical spine. Neck muscles can be divided according to their location in three regions: prevertebral region, lateral and anterior neck region or hyoid region. [28]

The region contains eight hyoid muscles divided into two groups: upper or suprahyoid and lower or infrahyoid.

The suprahyoid muscles, which are located over the hyoid bone, between it and the bony structures of the anterior region of the head, consist of 4 pairs of muscles: digastric, stylohyoid, mylohyoid and geniohyoid. The first two are on a superficial level and the last two on a deeper level. The main functions of this muscle group are lowering the jaw during mastication and phonetic processes, and the fixation of the hyoid bone to facilitate the action of the suprahyoid muscles in the swallowing process.

The digastric muscle extends from the base of the skull to the hyoid bone and from there to the central portion of the mandible. As its name suggests, it consists of two parts or bellies, one anterior and one posterior, united by an intermediate tendon. The anterior belly is oriented from back to front and from lateral to medial, toward the lower border of the mandible, a little outside the symphysis, in a special fossa called the digastric fossa. The contraction of the anterior belly, with a fixed point in the hyoid, directs the chin down and back (retropulsor and depressor of the jaw). In the contrary, if it takes its fulcrum in the digastric pit, it is a hyoid elevator.

The stylohyoid is a very thin, fusiform muscle, extending obliquely from the styloid process to the hyoid bone, inside and above the posterior belly of the digastric. It is a hyoid elevator.

The mylohyoid is a flattened and irregularly quadrilateral muscle that forms the mouth's floor. It is situated above the anterior belly of the digastric. It originates from fibers over the whole length of the ridge line or internal oblique or mylohyoid jaw. From there, their beams are directed downward and inward toward the midline and inserted, later, in the front of the hyoid; and earlier, in a central fascial raphe (suprahyoid white line), which extends from the inner surface of the chin to the hyoid body on its front face. The back of the muscle attaches to the body of the hyoid. The trailing edge is free at the distal face of the 3rd molar. With the jaw immobilized by other muscles, it elevates the hyoid with the larynx and moves the tongue against the palatal vault, playing an important role in the first half of swallowing. If the fulcrum is the hyoid, it is a depressor and in a lesser degree, a retropulsor of the jaw and it contributes to the opening of the mouth.

The geniohyoid is a small, cylindrical muscle located above the mylohyoid muscle, extending right and left toward the midline, from the hyoid to the symphysis of the chin. Its origin is in the lower genian apophysis and it inserts into the anterior surface of the hyoid bone. It has a dual action: in contraction, produces elevation of the hyoid bone, if it has a fixed point in the maxilla. It depresses the jaw if taken from a fixed point in the hyoid, previously immobilized by the contraction of the depressor muscles. [27,29,30]

1.12 Problem

Patients with Pierre Robin sequence present microretrognathia and growth studies show that patients rarely reach a normal development and mandibular growth although at early ages bone elongations and large overcorrections are made. Because of this abnormal mandibular growth, it is thought that the suprahyoid muscles are shorter and their function play a key role in the growth of the mandible.

1.13 Hypothesis

If patients with Pierre Robin sequence have micrognathia, then the suprahyoid muscles must be shorter and their function affects the normal development of the jaw.

1.14 General Objective

Using tomographic images, make longitudinal measurements of the muscles in the suprahyoid region in patients with Pierre Robin sequence and compare these measures with age-matched patients without this abnormality.

1.15 Specific Objective

Measure the length of the anterior belly of the digastric, mylohyoid and geniohyoid muscles in patients with Pierre Robin sequence and age-matched patients without this anomaly and compare results.

1.16 Material and Methods

1.16.1 Study universe.

35 patients with Pierre Robin sequence were treated in the Craniofacial Surgery Clinic of the Plastic and Reconstructive Surgery Department in the "Dr. Manuel Gea González" General Hospital during the period between June 2004 and December 2010. Of these patients, with or without surgical treatment, 15 underwent head and neck tomographic study. These studies were analyzed using MIMICS v program. 8.11 (The next generation for Intel X86 Platform V8.5.1.1.1 , 2004)

Patients were divided into 4 groups according to their age: Group 1 aged between 0 and 2 years; Group 2, aged 3 to 5 years; Group 3 ages from 6 to 9 years; Group 4, aged 10 to 12 years.

Simultaneously, 14 studies of children with similar ages and who featured a normal sized jaw were selected from the Department's file scans.

The measurements of the suprahyoid muscles were made in all the CT scans using sagittal projections for the anterior belly of the digastric muscle and cross section projections for the geniohyoid and mylohyoid muscles.

The anterior digastric was measured taking for reference the intermediate tendon of the digastric (located in the upper anterolateral face of the hyoid and the lower border of the mandible, a little outward of the symphysis, in the digastric fossa).

The mylohyoid was measured in the right and left side taking for reference the anterior surface of the hyoid and the mylohyoid ridge of the mandible. The measurement was taken in the middle half of the muscle (between the mandibular symphysis and the 3rd molar).

The geniohyoid was measured in the right and left side from the front of the hyoid bone to the inferior genial processes in the symphysis of the chin.

This is a descriptive, retrospective and comparative study.

1.16.2 Selection criteria:

Inclusion Criteria.

Patients included in the study had Pierre Robin sequence, were in a growing stage, and had a head and neck computed tomography scan, feasible for analyzing.

Exclusion criteria.

Adult patients with Pierre Robin sequence and those without an adequate head and neck computed tomography study.

Elimination criteria.

Poor definition of the structures to analyze in the tomographic study (bone and muscle references). Patients with classic mandibular osteotomies which modified the dimensions of the mandibular bone structure.

1.16.3 Variables.

Independent. Patient age. Dependent. Not applicable.

1.17 Ethical Considerations

All procedures were in accordance with the provisions of the Regulations of the General Health Law Responsible for Health Research. Title II, Chapter I, Article 17, Section I, in which no risk research does not require informed consent.

2 Preliminary Notes

2.1 STATISTICAL METHODS VALIDATION

For statistical analysis the SPSS v.16.0 software was used. For bivariate analysis, the Mann-Whitney U test was used (also called the Mann-Whitney-Wilcoxon) since it is the adequate statistic test for two ordinal heterogeneous samples such as the present work. Formulas:

$$U_1 = R_1 - \frac{n_1(n_1 + 1)}{2} \quad U_2 = R_2 - \frac{n_2(n_2 + 1)}{2}$$

3 Main Results

During the period from June 2004 to December 2010 there were 35 patients with Pierre Robin sequence treated at the Craniofacial Surgery Clinic of the Plastic and Reconstructive Surgery Department in the "Dr. Manuel Gea González" General Hospital". Of this group, 15 patients underwent a tomographic study of the head and neck. Finally, only 7 of these studies had the adequate technical characteristics to be analyzed with the MIMICS software version 8.11 (The next generation for Intel X86 Platform V8.5.1.1.1, 2004). The ages of the patients ranged from 1 to 12 years.

The measurements found in the Pierre Robin sequence patient group are shown in Table 1, which highlights the measurements in millimeters of the geniohyoid muscle among children 0-2 years and those aged 10 and 12, with ranges from 20.56 to 40.79 mm (average 29.11 mm). The dimensions of the anterior belly of the digastric when compared to the same age groups range from 18.58 to 26.31 mm (mean 22.06 mm). (Figure 1-A, B, C)

In contrast, measures of the control group patients also demonstrated a greater range of geniohyoid muscle growth, ranging from 22.64 to 44.24 mm (mean 32.11 mm) when comparing the lower age group with the highest. Also, in the control group, the second muscle showing major changes was the anterior digastric, measuring from 19.02 to 35.42 mm (mean 26.34 mm). (Table 2) (Figure 2-A, B, C).

In the statistical analysis, performed with SPSS v program. 16.0, univariate analysis results are reported in a histogram (Figure 3). Due to the small sample size, variables were not normally distributed (Kolmogorov-Smirnov), so the bivariate analysis was performed using a Mann Whitney test. As for the quantitative variables, the only one that presented normality was age, so the Student t test was used for finding differences in age between the healthy and Pierre Robin groups, not finding difference between them $p=0.628$. The rest showed abnormal distribution so nonparametric tests were performed:

Mann-Whitney U to compare variables (Table 3). No significant differences were found in any of the muscle groups.

4 Discussion

Pierre Robin sequence causes respiratory and digestive disorders, highlighting respiratory problems because the muscle mass of the floor of the mouth and the anterior volume of the neck occupy retropharyngeal space decreasing airway space.

Various techniques have been used for early treatment. In severe cases, it was very popular to use tracheostomy and muscle attachments of the tongue. The current trend is to solve by bone distraction. This technique has the additional advantage that it expands the overlying muscles of the bone structure that is elongated, it redistributes soft tissue of the floor of the mouth and the retropharyngeal space as well as it significantly increases the size of the airway, allowing children to breathe in a supine position and achieve normal oxygen concentrations in blood during sleep. However, although these children are overly corrected in the length of the jaw at an early stage, resulting in prognathous children, long term monitoring has shown that the jaw will not grow satisfactorily. Orthodontics, with the use of myofunctional apparatus are also used to stimulate growth of the facial bone structure in these patients.

Nevertheless, the long-term clinical follow-up has shown that all these children develop secondarily a class II molar bite and even again in many cases, micrognathia, where once again, the airway and food swallowing is compromised. This situation makes a reoperation necessary, for functional purposes as well as for aesthetics.

This study assesses the dimensions of the suprahyoid muscles and the results show that of this muscle group, the fastest growing muscle in children with normal jaw, but also in children with Pierre Robin sequence is the geniohyoid. Comparing the growth of this muscle in both groups, the difference ranges from 5.7 to 7.1 mm (mean 6.44 mm). (Table 4).

The most interesting finding corresponds to the anterior belly of the digastric as in the group of normal jaw, the growth between the lowest and highest age group was greater than 12 mm. However, comparing this muscle at the same ages, but with children with Pierre Robin, the maximum growth barely reached 6 mm. This fact explains the important natural tendency toward micrognathia and microgenia observed in these patients during their development and through adolescence. (Figure 4-A, B, C)

Finally, when comparing the mylohyoid muscle between the two groups, we found only millimetric differences, but in the morphological analysis of the growth of the mandibular body, the contraction of this muscle probably explains the slight concavity of the body of the jaws of the patients with Pierre Robin sequence unlike the slight convexity found in normal jaws. (Figure 5-A, B)

The posterior belly of the digastric muscle and the stylohyoid were not included because their contractility alter the mastoid and styloid apophysis respectively. These bony structures are separated from the jaw and we consider their dynamics do not affect the jaw's growth.

The comparative study of the suprahyoid muscles in children with Pierre Robin sequence and children with normal jaws corroborates the importance of the principle of functional matrix. In it, the continuous and prolonged interaction of different muscle groups with the

craniofacial skeleton significantly modify the bone structures in volume and dimensions and also secondarily affect their growth and development.

4.1 Advantages

Pierre Robin sequence is a widely studied clinical condition and patients with this condition are frequent in the Craniofacial Surgery Clinic of the Plastic and Reconstructive Surgery Department in the "Dr. Manuel Gea González" General Hospital. Patients are treated with different surgical procedures, however, after observation in a medium and long term, we can often observe a tendency to mandibular growth failure, so the study of the dimensions of the suprahyoid group of muscles that interact directly with the mandibular bone structure is necessary, in addition that medical literature has not reported any quantitative study of the extent to which the suprahyoid muscles are shortened compared with people without this condition.

4.1.1 Cost

The study didn't have any cost since the patients' tomographic studies and clinical photographs were taken from the archives, and the MIMICS program had already been acquired.

4 Labels of Figures and Tables

Table 1: Suprahyoid muscle measurements in patients with Pierre Robin sequence.
MYLO=MYLOHYOID, GENI=GENIHYOID

Age Group (YEARS)	Sex	Age	DIGASTRIC ANT BELLY (MM)	MYLO RIGHT (MM)	MYLO LEFT (MM)	GENI RIGHT (MM)	GENI LEFT (MM)
0 a 2	M	1	20.1	17.48	18.63	20.56	20.47
	F	2	18.58	22.9	22.39	22.7	22.82
3 a 5	F	3	19.14	21.91	19.93	32.47	30.99
	M	4	26.31	23.67	26.64	26.19	26.68
	F	5	19.37	19.04	17.65	29.9	29.84
10 a 12	F	11	25.6	24.06	23.15	32.15	33.04
	F	12	25.38	25.91	26.66	39.05	40.79

Table 2: Suprahyoid muscle measurements in patients with normal jaw.

MYLO=MYLOHYOID, GENI=GENIHYOID

Age Group (YEARS)	Sex	Age	DIGASTRIC ANT BELLY (MM)	MYLO RIGHT (MM)	MYLO LEFT (MM)	GENI RIGHT (MM)	GENI LEFT (MM)
0 a 2	M	1	20.05	18.93	19	22.64	22.65
	M	1	21.78	18.81	19.43	23.46	23.74
	M	1	23.8	21.67	21.05	26.18	27.11
	F	2	23.07	23.76	23.34	28.53	28.76
	F	2	19.02	22.3	20.75	29.76	30.04
	M	2	24.11	22.34	21.45	33.43	34.18
3 a 5	F	3	25.2	22.11	21.85	33.54	33.3
	M	4	27.49	25.07	26.79	31.29	31.42
	F	4	28.5	27.03	25.78	28.14	28.41
	M	4	27.67	24.06	25.12	29.27	29.47
	F	5	24.64	20.48	22.26	32.47	32.91
10 a 12	M	10	35.42	26.79	27.55	40.78	41.37
	F	12	35.25	30.82	28.53	43.33	43.91
	M	12	32.89	28.56	28.47	44.24	44.8

Table 3: Bivariate analysis Report with non-parametric Mann Whitney U test where no significant differences were found in any of the muscles.

Test Statistics^a

	DIGASTRIC ANT BELLY	MYLOHYOID RIGHT	MYLOHYOID LEFT	GENIOHYOID RIGHT	GENIOHYOID LEFT
Mann-Whitney U	28.000	38.500	36.000	38.500	36.000
Wilcoxon W	56.000	66.500	64.000	66.500	64.000
Z	-1.567	-.784	-.970	-.784	-.970
Asymptotic difference (two-tailed)	.117	.433	.332	.433	.332

a. Grouping Variable: Pierre Robin

Table 4: Millimetric difference in muscle length between patients with Pierre Robin sequence and patients with normal jaw. MYLO=MYLOHYOID, GENI=GENIHYOID

Age Group (YEARS)	DIGASTRIC ANT BELLY (MM)	MYLO RIGHT (MM)	MYLO LEFT (MM)	GENI RIGHT (MM)	GENI LEFT (MM)
0 a 2	2.631666667	1.111667	0.326667	5.703333	6.101667
3 a 5	5.093333333	2.21	2.953333	1.422	1.932
6 a 9	0	0	0	0	0
10 a 12	9.03	3.738333	3.278333	7.183333	6.445



Figure 1-A: Twelve year old female patient with Pierre Robin sequence where the length of the anterior belly of the digastric was measured from the digastric fossa in the posterior region of the chin to the front of the hyoid body.

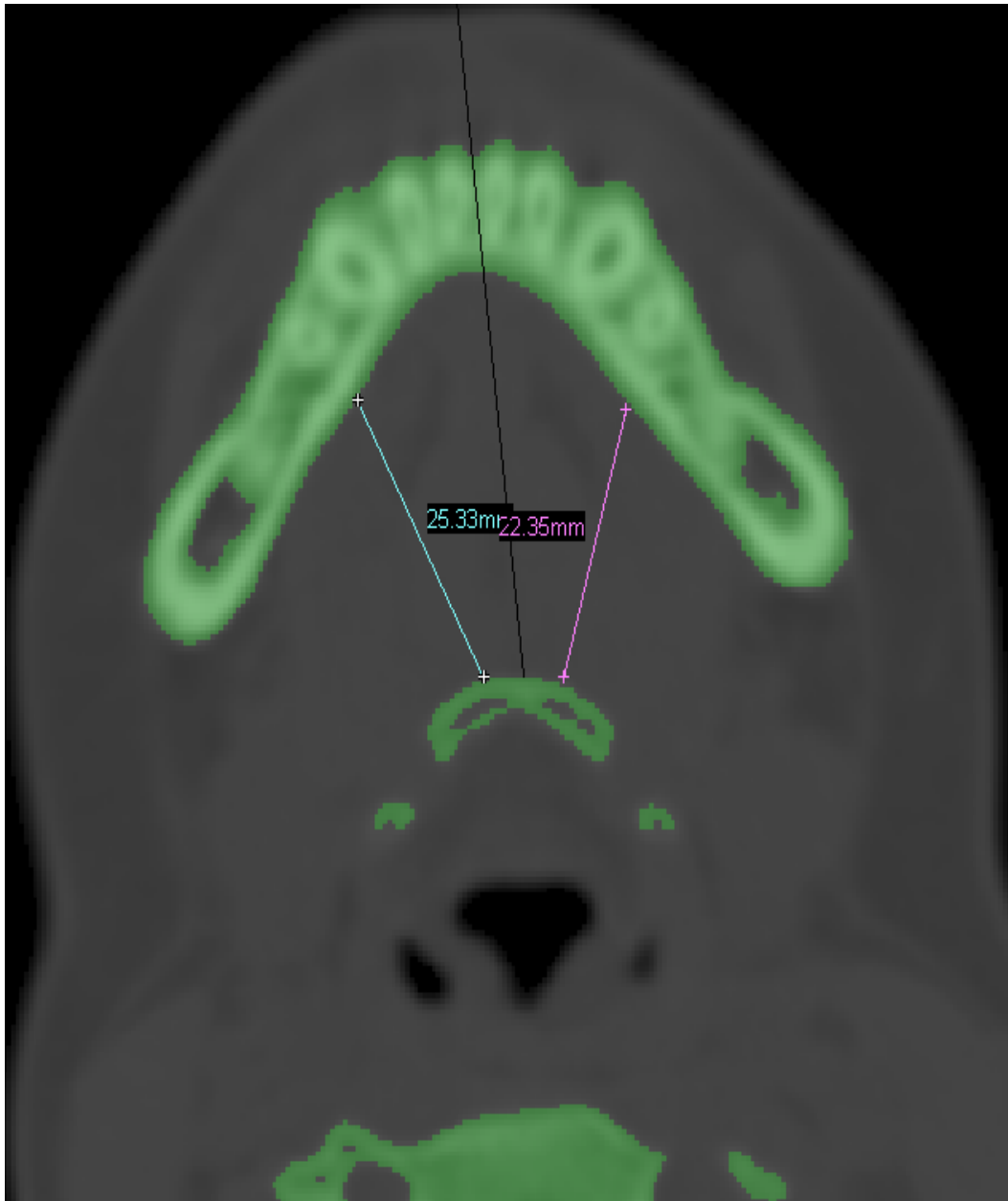


Figure 1-B: Twelve year old female patient with Pierre Robin sequence where the length of the mylohyoid muscles were measured from the front of the hyoid to the mylohyoid ridge of the mandible (between the mandibular symphysis and the 3rd molar).

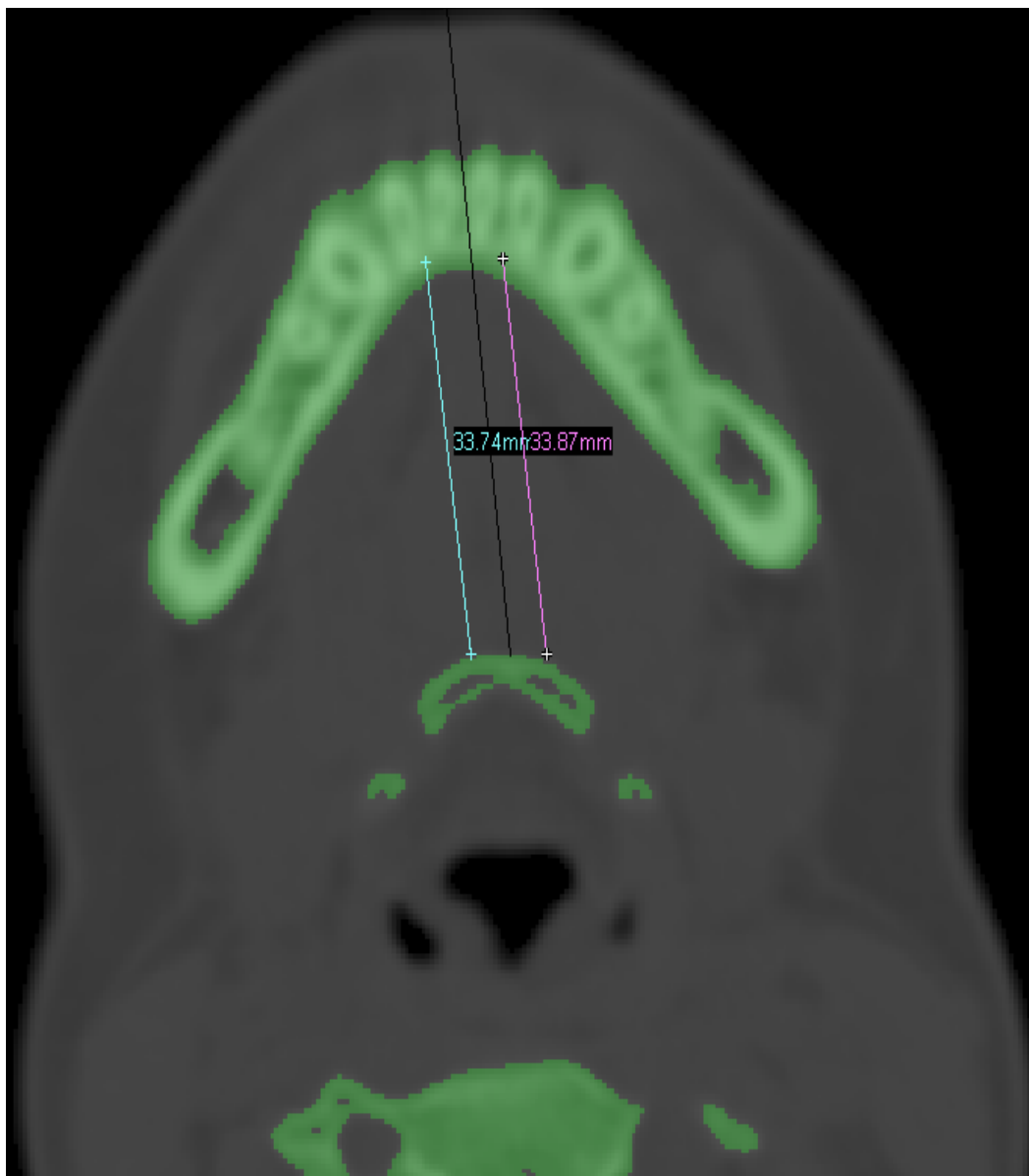


Figure 1-C: Twelve year old female patient with Pierre Robin sequence where the length of the genioid muscles were measured from the anterior surface of the hyoid to the lower genial processes in the posterior region of the chin.

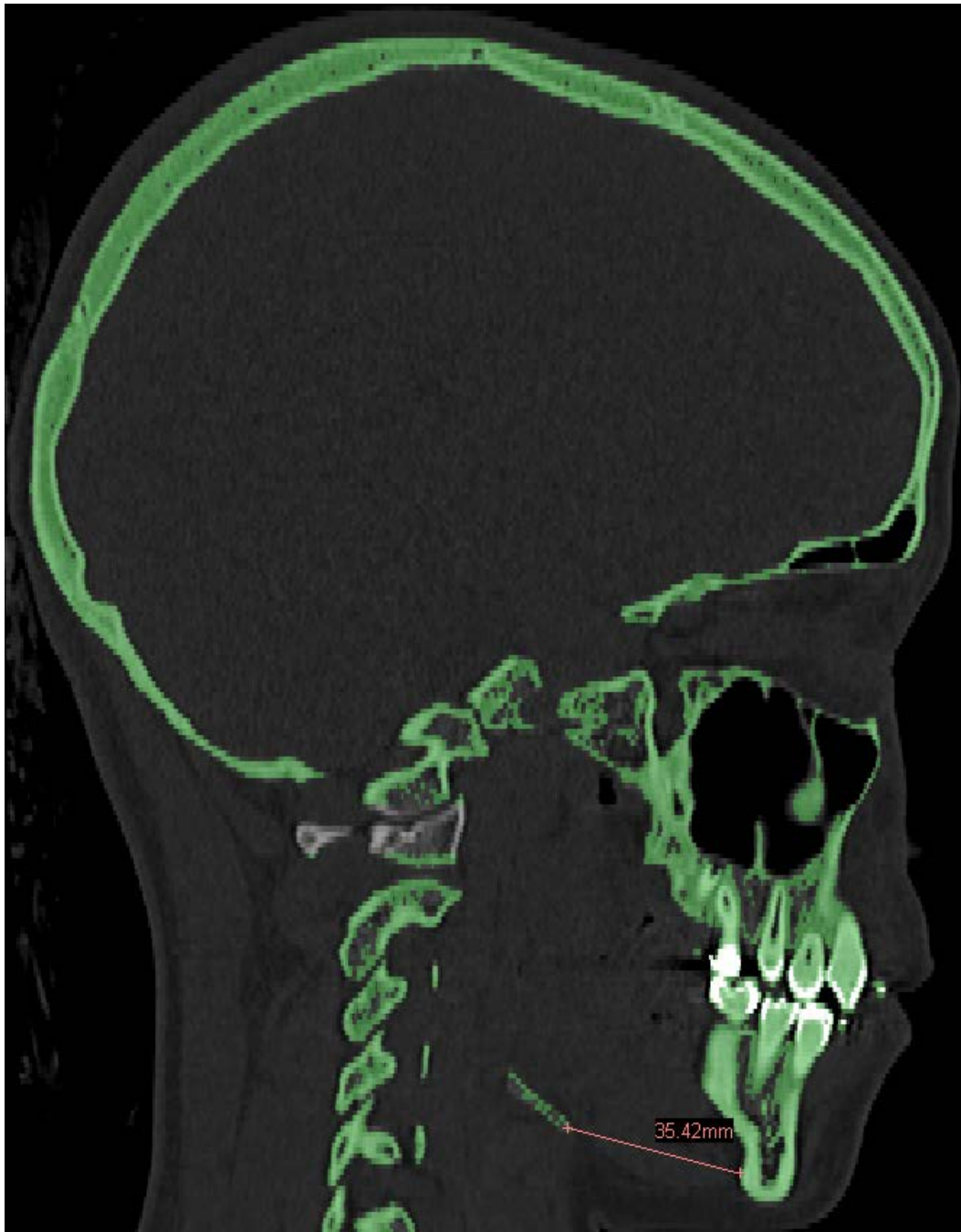


Figure 2-A: Twelve year old female patient with normal jaw where the length of the anterior belly of the digastric was measured from the digastric fossa in the posterior region of the chin to the front of the hyoid body.

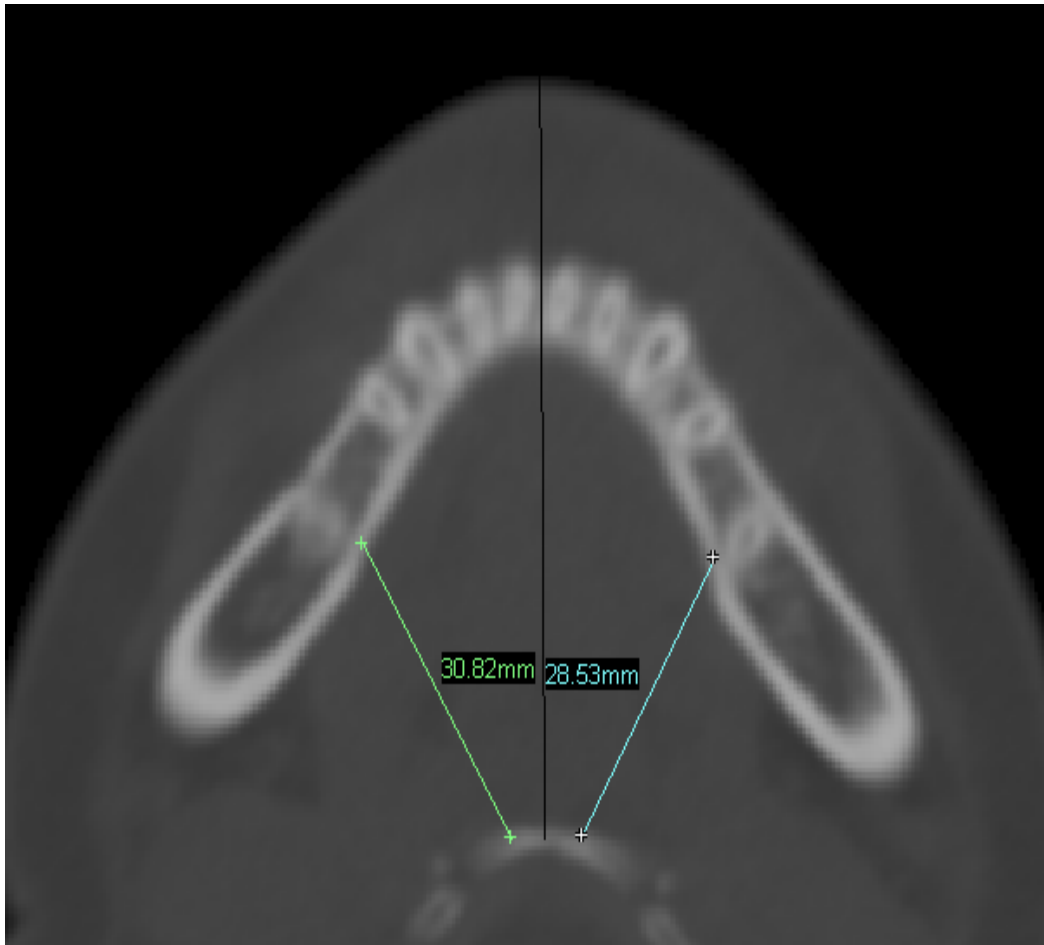


Figure 2-B: Twelve year old female patient with normal jaw were the length of the mylohyoid muscles were measured from the front of the hyoid to the mylohyoid ridge of the mandible.



Figure 2-C: Twelve year old female patient with normal jaw were the length of the geniohyoid muscles were measured from the anterior surface of the hyoid to the lower genial processes in the posterior region of the chin.

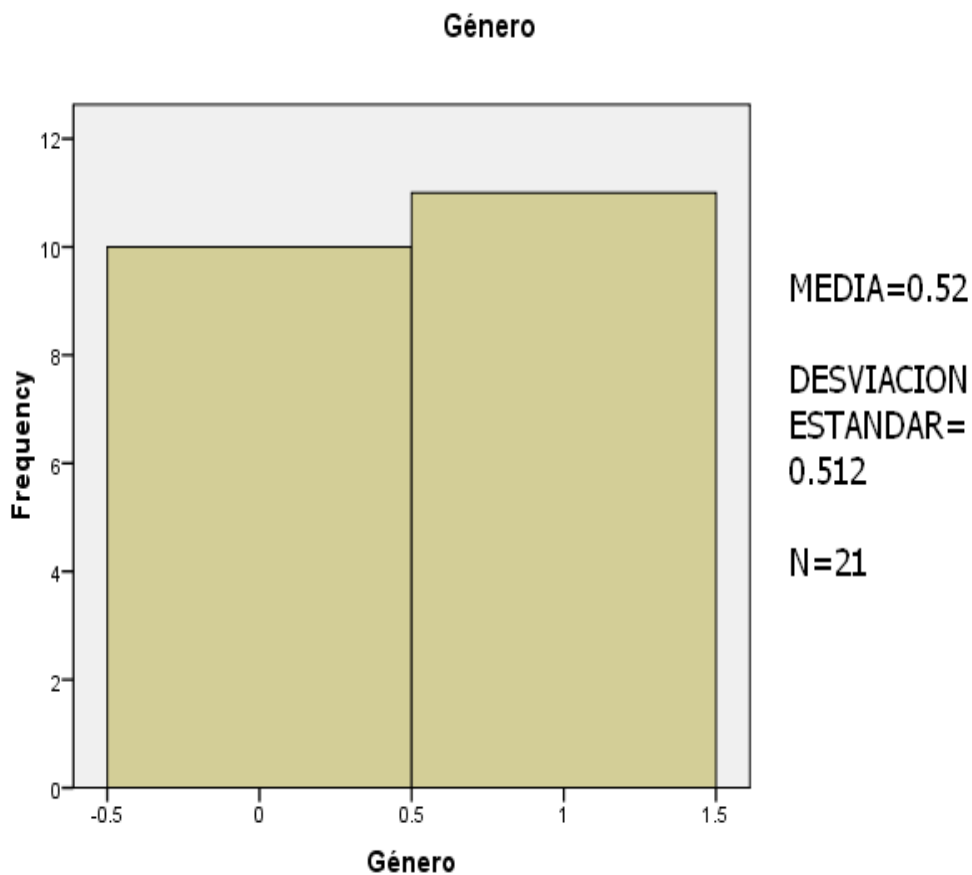


Figure 3: Histogram showing the results of the univariate analysis for categorical variables frequency.



Figure 4-A: Two year old female patient with Pierre Robin sequence.



Figure4-B: Same patient at age 4, after mandibular distraction and over-correction.



Figure4-C: Same patient at age 12, showing severe severe microgenia and decreased size of the mandibular body. Analyzing the occlusion she presents a typical Angle class II.

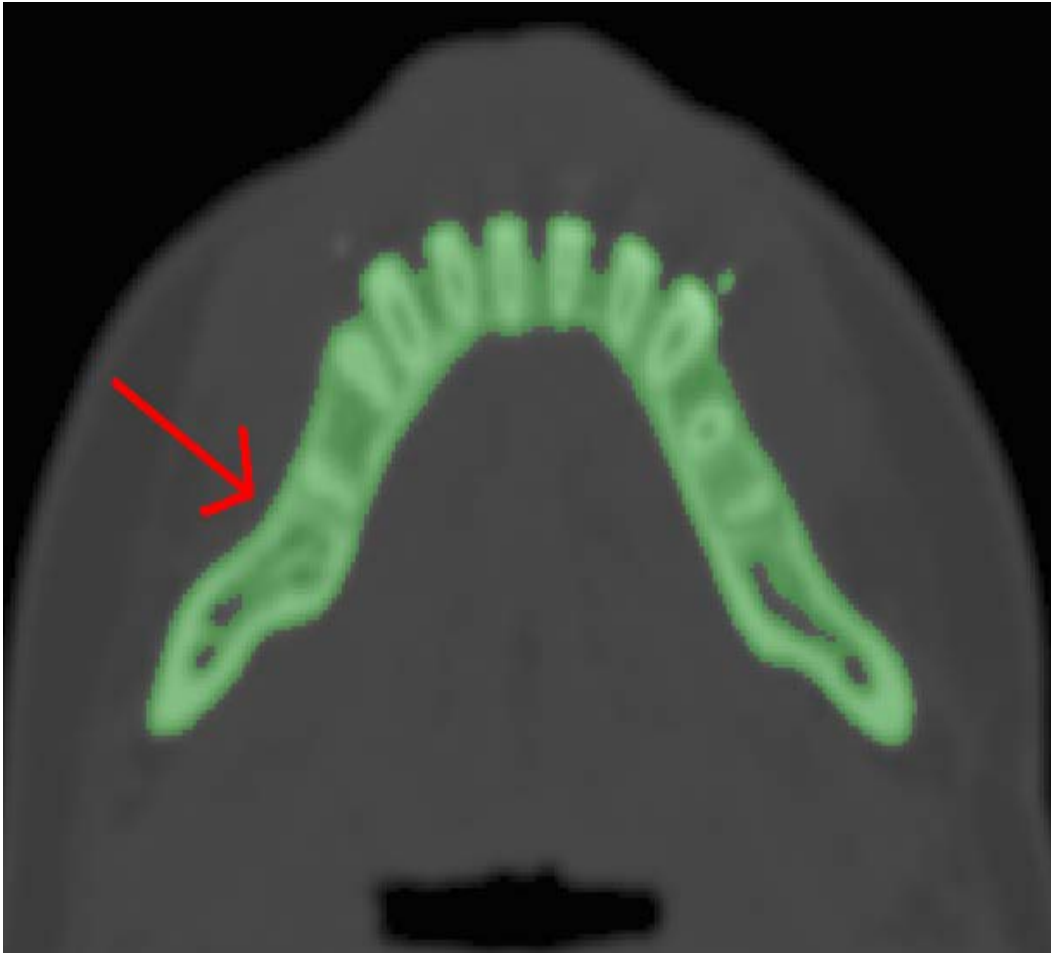


Figure5-A: Cross section of the mandible of a patient with Pierre Robin sequence. The shortness of the mylohyoid muscle conditions a discrete concavity in the mandibular body.

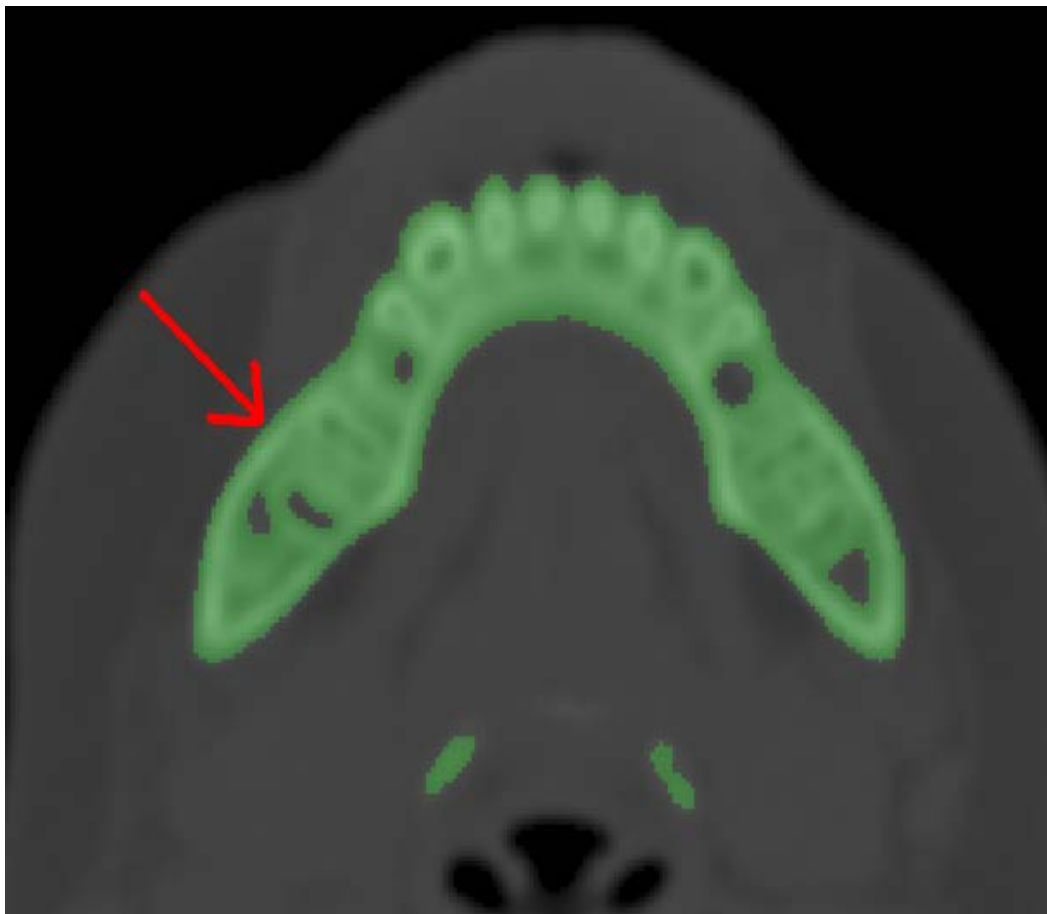


Figure5-B: Cross section of a normal jaw in a 3 year old patient. Typical dimensions of the mylohyoid muscle fibers produce a discrete convexity on the jaw body.

5 Conclusion

Pierre Robin sequence is a disorder characterized by micrognathia and patients often suffer severe respiratory problems since birth.

The surgical techniques used in its treatment satisfactorily resolved impaired respiratory function, however, the natural tendency of the disease is that the jaw does not grow satisfactorily and in puberty and adolescence the patients have microgenia and micrognathia. This study demonstrates the important role of the suprahyoid muscle group, which through its short length and contractile function secondarily affect the growth of the mandible.

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