# Macrodontia, Shovel-shaped Incisors, and Multituberculism: Probable Ekman-Westborg-Julin Trait

*Gayle Tieszen Reardon, DDS, MA, MS*
*Rebecca L. Slayton, DDS, PhD*
*Clinton Norby, DDS*
*Teresa Geneser, DMD*

## ABSTRACT

Multiple macrodontia is a rare finding and is defined as a condition in which a tooth is significantly larger than normal. Macrodontia may occur as an isolated finding, part of a group of dental anomalies, or as a component of a syndrome with multiple oral and systemic manifestations. The purpose of this paper was to report a case of macrodontia affecting all permanent teeth and exhibiting shovel-shaped maxillary and mandibular incisors and multituberculate molars and premolars. Some or all of this patient's characteristics have been reported in both males and females, with a ratio of 5:2. No inheritance pattern has been established, as these traits have generally occurred spontaneously. As more individuals are identified and as molecular techniques continue to advance, it is probable that a gene or genes responsible for macrodontia and the associated traits will be identified. (J Dent Child 2012;79(3):197-201)

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Anomalies in tooth development are relatively common and affect the size, shape, number, and mineralization of the teeth. Some dental anomalies are seen as sporadic findings, while others are associated with syndromes. Multiple anomalies in one individual are suggestive of a systemic cause, such as a genetic mutation.

Macrodontia is a term used to describe the presence of 1 or more teeth that are larger than normal.[1] Normal tooth dimensions vary by gender and racial background. G.V. Black published early studies of normal tooth sizes in his textbook "Descriptive Anatomy of the Human Teeth" in 1897.[2] Macrodontia may occur as an isolated finding or part of a syndrome or condition with multiple oral and systemic manifestations such as KBG syndrome and congenital hemifacial microsomia.[1] The most commonly affected teeth are the second premolars and third molars,[3] while incisors are rarely affected. In a study of 3,043 Turkish children undergoing orthodontics, there was only 1 child (<1%) with macrodontia of a single tooth.[4] Generalized macrodontia is extremely rare, with only a few reported cases in the past 30 years.[4-8] Generalized macrodontia is also seen in cases of pituitary gigantism.[1] Shovel-shaped incisors are relatively common among individuals of Asian and Native American heritage.[9] Inheritance of this morphologic trait is thought to be polygenic. Recently, an association between polymorphism in the ectodysplasin A receptor gene and the presence of incisor shoveling was found in 2 Japanese populations.[9] While this polymorphism does not explain all occurrences of incisor shoveling, it suggests that genetics plays an important role in the etiology of this dental anomaly.

Multiple dental anomalies in one individual are rare and suggest an underlying genetic etiology. Ekman-Westborg and Julin described a 14-year-old patient

*Dr. Reardon was a resident in Oral Radiology at the University of Iowa College of Dentistry and is now in private practice in Sioux Falls, South Dakota, Dr. Slayton is professor and chair, Department of Pediatric Dentistry, University of Iowa College of Dentistry, Iowa City, Iowa, Dr. Norby was a resident in OMFS at the College of Dentistry and is now in private practice in Dakota Dunes, S.D. and Dr. Geneser was a resident in OMFS at the Iowa College of Dentistry and is now in private practice in Coralville, Iowa.*
*Correspond with Dr. Reardon at gtrdds@aol.com*

with multiple dental anomalies, including multiple macrodonts, impacted teeth, and posterior crossbite.[5] The child's hair was noted to be very coarse and curly compared to the hair of other family members. One maxillary canine had a peak-shaped cusp with a step on the mesial side, suggesting a separate mesial cusp. The crowns of premolars and molars were notably "larger than normal" and exhibited several extra cusps. A radiographic examination of the same patient exhibited third molars more than twice their normal size, with multiple cusps mimicking the form of a walnut kernel. Roots were not present. The premolars evidenced both invaginations and evaginations.

Multiple macrodonts with giant third molars, combined with multituberculism, structural invaginations and evaginations, and peak-shaped canines, came to be called Ekman-Westborg-Julin trait (**EWJT**).[6] Since the original paper[5], multiple reports have described similar findings in both male and female subjects.[6-8,10,11] Nemes and Alberth[6] described a 7-year-old girl with generalized macrodontia of the permanent teeth, shovel-shaped maxillary incisor crowns, multiple molar cusps, and invaginations on all maxillary incisors and premolars. Benjamin et al.,[7] reported a case of a 12-year-old Chilean girl with shovel-shaped maxillary and mandibular incisors, premolars with evaginations and multiple tubercles, and macrodontic molars with multiple tubercles. The molar roots were thin and the maxillary molar roots were conical in shape. The authors argued that these features, although consistent with those previously reported for EWJT, did not fit the definition of a syndrome and should be referred to as multiple macrodontic multituberculism. By definition, a syndrome is a "set of symptoms which occur together."[12]

## CASE REPORT

A 12-year, 2-month old male presented to the pediatric dentistry clinic of the College of Dentistry, University of Iowa, Iowa City, Iowa, for evaluation. He was referred by the family's general dentist who was concerned about "multiple macrodontic teeth and the failed eruption of multiple teeth." The patient, his father and older brother, were originally from Guatemala. The father reported that he had attempted to get dental treatment for his son in the past, but previous dentists were not sure how to manage his care. At this visit, the patient had no complaint of pain but was concerned about the appearance of his teeth.

The patient's medical history included tonsillectomy, myringotomy tubes, nasal surgery due to snoring and mouth-breathing, and a learning disability. An oral examination was completed by a pediatric dentistry resident, and consultations were requested with the Departments of Radiology, Prosthodontics, Orthodontics, and Oral Surgery at the University of Iowa College of Dentistry.

An extraoral examination disclosed no edema, erythema, or tenderness on palpation. The face appeared dolicocephalic and the patient had hypertelorism and a broad nasal bridge. The intraoral examination revealed that his permanent maxillary and mandibular incisors were macrodonts which were shovel-shaped and had

*Figure 1. Mandibular arch showing erupted shovel-shaped incisors and canines with invaginations and evaginations.*

*Figure 2. Maxillary arch at the initial exam showing steep palatal vault, carious premolars, shovel-shaped incisors, and multituberculate molars and premolars.*

*Figure 3. Frontal view at the initial exam showing excess crowding in the maxillary arch, a labially positioned maxillary right central incisor, ectopically erupting maxillary left canine, rightposterior crossbite, plaque, and gingivitis.*

evaginations on their lingual surfaces (Figures 1-2). The mandibular canines had invaginations and central evaginations (Figure 1). The permanent maxillary left canine was partially erupted and peak-shaped with an extra mesial cusp, but the permanent maxillary right canine was unerupted (Figures 2-3). The permanent maxillary first premolars were macrodonts with occlusal caries, invaginations, and evaginations (Figure 2). The erupted permanent maxillary molars were multitubercle macrodonts with crenulated occlusal surfaces (Figure 2). The permanent maxillary left central incisor was clinically missing due to avulsion secondary to trauma, which had occurred a few years earlier (Figure 3).

The palatal vault was very steep, with thickened alveolar ridges (Figure 2). All mandibular premolars and molars were impacted, and there was significant crowding in both arches due to a tooth size/arch length discrepancy (Figure 4). The patient's oral hygiene was poor,

***Table 1. Mesiodistal and Buccolingual Dimensions of Extracted Macrodonts and Normal Teeth[2]***

| Tooth type (no.) | Mesiodistal dimension (mm) | | Buccolingual dimension (mm) | |
|---|---|---|---|---|
| | Macrodont | Normal | Macrodont | Normal[2] |
| Maxillary central incisor (8) | 11 | 9 | 7.5 | 7 |
| Maxillary first premolar (5) | 12 | 7.2 | 10 | 9.1 |
| Maxillary first premolar (12) | 11 | 7.2 | 12 | 9.1 |
| Maxillary second premolar (4) | 14.5 | 6.8 | 13.5 | 8.8 |
| Maxillary second premolar (13) | 15 | 6.8 | 12 | 8.8 |
| Maxillary second molar (2) | 16 | 9.2 | 16 | 11.5 |
| Maxillary second molar (15) | 15.5 | 9.2 | 16 | 11.5 |
| Maxillary third molar (1) | 21.5 | 8.6 | 15 | 10.6 |
| Maxillary third molar (16) | 20 | 8.6 | 15 | 10.6 |

*Figure 4. Preoperative panoramic radiograph showing multiple mandibular impacted macrodonts and a maxillary left peridens (supernumerary tooth) between teeth nos. 14 and 15.*


*Figure 5. Panoramic radiograph, taken immediately postoperative, showing remaining teeth and areas where bone was removed to access the impacted teeth.*

*Figure 6. Frontal view 6 months after surgery.*

*Figure 7. Maxillary occlusal view 6 months after surgery.*

with plaque evident on all tooth surfaces, thus producing a moderate level of generalized gingivitis. The maxillary central incisor was labially positioned, and the maxillary left canine was erupting ectopically due to severe crowding (Figures 3-4). There was also a right posterior crossbite.

A panoramic radiograph exhibited multiple macrodont teeth, impacted mandibular and maxillary posterior teeth, and mixed radiopaque tooth-like structures associated with the unerupted mandibular third molars, which were positioned in the angle regions of the mandibular rami bilaterally (Figure 4). The unerupted mandibular teeth were situated very near the inferior border of the mandible. In the maxillary left posterior quadrant, there was a peridens (supernumerary tooth) positioned between the permanent first and second molars.

A cone beam computed tomography scan was ordered to further investigate the osseous, cervical spine, sinus, and dental regions of the head and neck and to determine the precise locations of the impacted teeth.

## TREATMENT

After consultation with the Departments of Prosthodontics and Orthodontics, the final treatment plan included extraction of all impacted molars and premolars and the restoration or possible extraction of the carious maxillary premolars. The solitary maxillary right central incisor was planned for extraction due to previous loss of its counterpart and the overall lack of space in the maxillary arch. Informed consent was obtained from the patient's father for surgical treatment under general anesthesia. The treatment was provided by the Oral and Maxillofacial Surgery Department in a sterile operating suite under general anesthesia.

In both maxillary posterior quadrants, a full thickness flap was raised and bone was removed to provide access to the impacted teeth. Teeth numbers 1, 2, 13, 14s (peridens), 15, and 16 were extracted with elevation. Teeth numbers 4 and 8 were removed using a simple forceps technique. All areas were curetted, rinsed, and then closed with gel foam and 3-0 chromic gut sutures. A pediatric dentist determined that teeth numbers 5 and 12 were non-restorable; they were extracted with a forceps. The sockets were curetted, rinsed, and then closed with gel foam and chromic gut sutures.

Teeth numbers 17, 18, 19, 20, 29, 30, 31 and 32 required surgical sectioning and removal in pieces. Teeth numbers 21 and 28 required bony removal and elevation. All sockets were curetted and rinsed. An alveoloplasty was performed adjacent to the sockets of teeth numbers 30 and 31. Primary closure was accomplished with 3-0 chromic gut sutures. The inferior alveolar nerve was not visualized on the left side but it could be seen on the right side and remained intact at the apex of tooth number 30.

The procedure was completed successfully, and the patient was transferred to the post-anesthesia care unit in good condition. Immediately following his release from second stage surgery, a follow-up panoramic radiograph was made to assess the integrity of the inferior border of the mandible and to confirm that no inadvertent fracture had occurred during the surgery (Figure 5). The postoperative instructions included a soft diet, chlorhexidine gluconate oral rinse, amoxicillin clavulanate for 7 days, and avoidance of contact sports for 3 months.

## HISTOPATHOLOGY

All examined teeth presented similar gross findings. All were larger than expected with enlarged coronal/incisal surfaces exhibiting supernumerary rudimentary cusps/tubercles (Table 1). The anterior teeth displayed irregular incisal edges, prominent and deformed cingula, and radicular malformation. One surgically divided molar tooth with sectioning through the pulpal chamber was further examined histopathologically. Minimal pulpal tissue and periodontal ligament remnants were grossly present. Following demineralization, the histopathology showed an abnormally shaped and sized tooth with generally normal dentin and dentinotubular structure, although the length of tubules was often greatly extended. There was some predentin present with focally prominent odontoblasts. The tooth was notable for very long, thin pulp horns extending for several millimeters from the main chamber and ending very close to the rudimentary cusps/tubercles.

## FOLLOW–UP

The patient was seen for a 2-week postoperative visit with the University of Iowa Department of Oral and Maxillofacial Surgery. At this visit, he had no complaints and healing was progressing well. It was noted that his oral hygiene was still poor and he was encouraged to improve the frequency and efficacy of his tooth-brushing.

Six months following his surgery, he was seen in the pediatric dentistry clinic for evaluation of healing, oral hygiene, and occlusion. His oral hygiene had improved considerably, his maxillary canines were erupting into place, and the lateral incisors were in the position of the missing central incisors (Figures 6-7). The partially erupted maxillary left canine displayed invaginations and evaginations of the palatal surface, similar to that seen with the mandibular canines. At this visit, resin sealants were placed to protect the occlusal surfaces of the molars and lingual surfaces of the incisors and canines from future caries. A 6-month recall visit with his local general dentist was recommended due to the families' long drive to the College of Dentistry.

In the future, this patient would benefit from orthodontics to expand his maxilla and to position his incisors and canines in a more ideal occlusal relationship. Once his growth is complete, evaluation for placement of dental implants in the posterior mandible should be considered to improve function and preserve alveolar bone.

## DISCUSSION

The dental features in this patient are consistent with the EWJT and multiple macrodontic multituberculism cases described previously.[4-8,11] Unique to this case were generalized macrodontia and the impaction of all mandibular and most maxillary molars. These and other findings such as the shovel-shaped incisors, multituberculism of molars and premolars, invaginations and evaginations of incisors, premolars, and canines, and intellectual developmental disability are consistent with other reports.[4-8, 11]

Although some of these features are consistent with KBG syndrome, there are not enough to make it the likely diagnosis. In KBG syndrome, the macrodontia is limited to the maxillary central incisors, whereas, in this case there was generalized macrodontia. KBG syndrome patients have characteristic facial features, including triangular facies, synophrys, brachycephaly, and a short webbed neck. Our patient was dolichocephalic, had sparse eyebrows, and lacked webbing of the neck. There was no report of neurological issues, seizures, or anxiety disorder; however, he had a mild learning disorder.

Shovel-shaped incisors, as isolated findings, are not uncommon among individuals of Asian and Native American heritage.[9] When comparing incisor shoveling among Asian and African populations, Kharat and colleagues found incidences ranging from 5% among Syrians, Jordanians, Palestinians, and Filipinos and 20% to 25% among Yemenis, Sudanese, and Egyptians.[13] Most published studies of incisor shoveling focus on maxillary incisors, with relatively few specifically mentioning the mandibular incisors.[14, 15] Since the adolescent in this case report was from Guatemala, where one of the more common ethnic groups is Amerindian, it is possible that the shovel-shaped incisors are more related to his ethnicity than the other dental anomalies he presented. Neither his father nor brother, however, had shovel-shaped incisors.

Varying combinations of the characteristics described by Ekman-Westborg and Julin in 1974 have been reported in both males and females, at a ratio of 5:2.[6] No inheritance pattern has been established, as these traits have generally occurred spontaneously. One case report of a mother and son suggested an autosomal dominant inheritance pattern.[11] No reported cases have involved parental consanguinity, and no cases have documented affected siblings. In the present case, the patient's father and older brother have complete permanent dentitions with normally shaped and sized teeth. His mother has not been in his life for many years and thus could not be questioned or examined. Based on his history, it is likely that these anomalies are due to a sporadic mutation.

The occurrence of these traits in both males and females suggests an autosomal dominant inheritance pattern. An autosomal recessive inheritance pattern, however, cannot be ruled out. None of the reported cases has included histories of grandparents or distant relatives. Genetic analysis is possible but challenging with so few cases to evaluate. To date, there have been no suggestions for a genetic cause of macrodontia. As more individuals are identified and molecular techniques continue to advance, it is probable that a gene or genes responsible for macrodontia and the associated traits will be identified.

## REFERENCES


1. Laskaris G. Color Atlas of Oral Diseases in Children and Adolescents. New York, NY: Thieme; 2000:2-4.
2. Black GV. Descriptive Anatomy of the Human Teeth. Philadelphia, PA: The S.S. White Dental Manufacturing Company; 1897:16-17.
3. Dugmore CR. Bilateral macrodontia of mandibular second premolars: A case report. Int J Paediatr Dent 2001;11:69-73.
4. Altug-Atac AT, Erdem D. Prevalence and distribution of dental anomalies in orthodontic patients. Am J Orthod Dentofacial Orthop 2007;131:510-4.
5. Ekman-Westborg B, Julin P. Multiple anomalies in dental morphology: Macrodontia, multituberculism, central cusps, and pulp invaginations: Report of a case. Oral Surg Oral Med Oral Pathol 1974;38:217-22.
6. Nemes JA, Alberth M. The Ekman-Westborg and Julin trait: Report of a case. Oral Surg Oral Med Oral Pathol Oral Radiol Endod 2006;102:659-62.
7. Benjamin Martinez R, Rodrigo Farina S, Gorlin, RJ. Clinical report: Multiple macrodontic multituberculism. Am J Med Genet 2003;120A:283-5.
8. Reichart PA, Triadan H, Lenz H. Polyanomalodontia. J Oral Pathol Med 1978;7:100-8.
9. Kimura R, Yamaguchi T, Takeda M, et al. A common variation in EDAR is a genetic determinant of shovel-shaped incisors. Am J Hum Genet 2009; 85:528-35.
10. Nakagawa T, Tagawa T, Inui M, Kageyama T, Murata T. Ekman-Westborg-Julin syndrome: A case report. Int J Oral Maxillofac Surg 1997;2:26:49-50.
11. Yoda T, Ishii Y, Honma Y, Sakai E, Enomoto S. Multiple macrodonts with odontoma in a mother and son: A variant of Ekman-Westborg-Julin syndrome. Oral Surg Oral Med Oral Pathol Oral Radiol Endod 1998;85:301-3.
12. Dorland's Illustrated Medical Dictionary, 32nd Edition. St. Louis, MO: Saunders; 2011.
13. Kharat DU, Saini TS, Mokeem S. Shovel-shaped incisors and associated invagination in some Asian and African populations. J Dent 1990;18:216-20.
14. Saini TS, Kharat DU, Mokeem S. Prevalence of shovel-shaped incisors in Saudi Arabian dental patients. Oral Surg Oral Med Oral Pathol 1990;70:540-4.
15. Hsu JW, Tsai P, Liu K, Ferguson D. Logistic analysis of shovel and Carabelli's tooth traits in a Caucasoid population. Forensic Sci Int Genet 1997;89:65-74.