Scleroderma: a case report of possible cause of restricted movement of the temporomandibular joint with effects on facial development

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Scleroderma is a rare abnormality in which fibrosis of the skin, subcutaneous tissues and muscle may occur. All forms of scleroderma are rare in childhood: the most common form is localized scleroderma. Localized scleroderma is often benign, but may cause significant deformity, if it occurs on the face or extends across joint surfaces. Structural changes may occur in the osseous tissue and result in mandibular joint restriction (pseudoankylosis) and facial and occlusal disharmonies. This article describes a case of facial linear scleroderma in a nine-year-old child and the treatment performed to minimize consequences on occlusion and face development. Available clinical data will be illustrated.
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INTRODUCTION

Rapid advances in immunology have contributed significantly to our understanding of immunopathogenesis and of immunologica diseases: the concept of many diseases considered to be of unknown etiology are changing drastically with these advances.

The basic function of the immune system is to distinguish self from non-self, thus eliminating the injurious and pathogenic potential of foreign substances introduced into the body. The immune response is a biological phenomenon necessary for protection, injury mediation and repair foundation. The initiation of this response is triggered by the presence of an antigen, which can combine with host proteins to initiate the immune response and antibody production. The autoimmune diseases are characterised as multisystem, inflammatory process-related diseases with a predisposition towards connective tissue and vascular structures that manifest an autoimmune abnormality. Autoimmunity is simply the production of antibodies that react with normal tissues in response to mutation or organism-tissue cross-reaction.

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Scleroderma is a rare abnormality[1] in which fibrosis of the skin, subcutaneous tissues and muscle may occur, not of necessity affecting the face and often associated with similar changes in the alimentary tract manifested by dysphagia attributed to pharyngeal sclerosis. It includes two diseases that are characterized by hardening and tightening of the skin, but appear to have different etiologies. Localized (linear) scleroderma is deforming, but not lethal or diffuse. Progressive systemic scleroderma involves the skin as well as internal organs. The linear scleroderma, characterized by a thin band of sclerosis covering the length of an extremity and involving underlying muscles, bones, and joints is common in childhood. When the face is involved, oral manifestations may include rigid lips, reduced mouth opening and tongue sclerosis, which may limit mandibular movements. Alteration in the occlusion of the upper and lower dentition may result in alteration of the position of the condyle in relation with the fossa. Besides, structural changes may occur in the osseous tissue of the face and mandible resulting in mandibular joint restriction (pseudoankylosis). Unfortunately scant reference to this is seen in literature.[2]

This article describes a case of facial linear scleroderma in a nine-year-old child and the treatment performed to minimize consequences on occlusion and facial development, so that others may evaluate it, and possibly benefit from our experience.

CASE REPORT

A nine-year-old girl was referred to by the pediatrician for clinical and radiolographic examination following a diagnosis of facial localized scleroderma (Figure 1). The

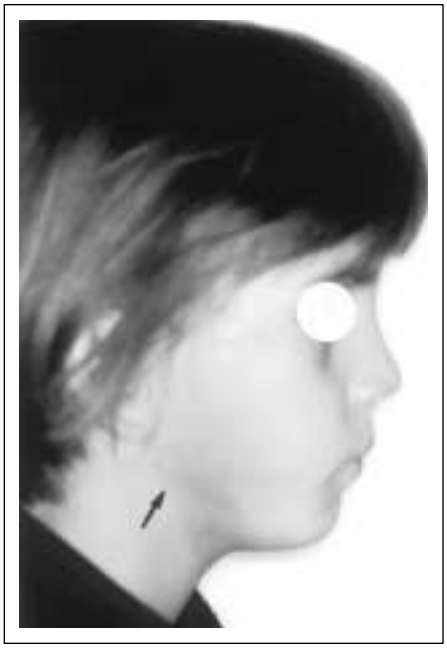


Figure 1. Lateral photograph: the skin lesion on the right cheek of the patient is evident.

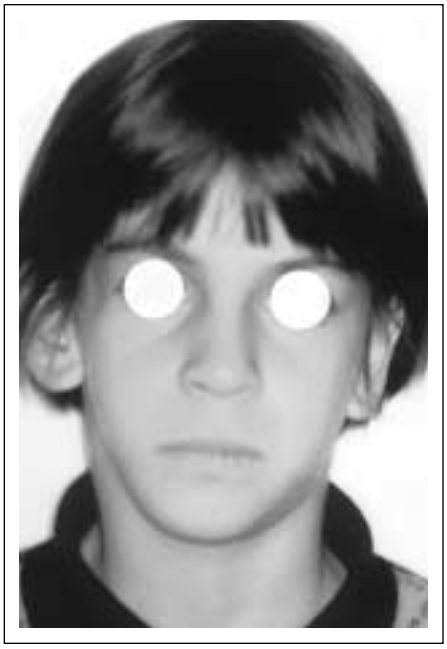


Figure 2. Frontal photograph: a light shift of the chin towards the right side and a deviation of the midline to the right are present.

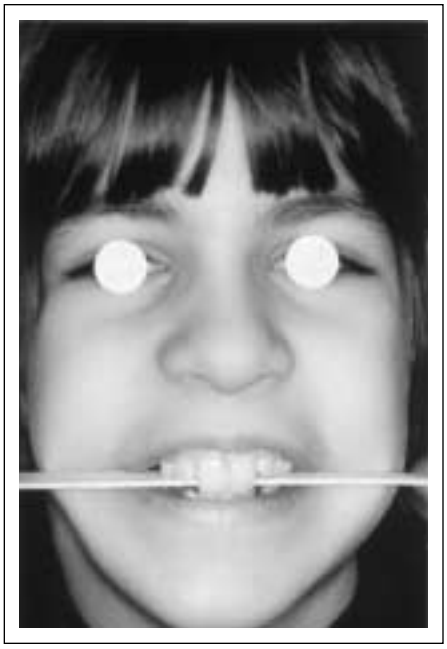


Figure 3. Frontal photograph: the occlusal plane is flat.

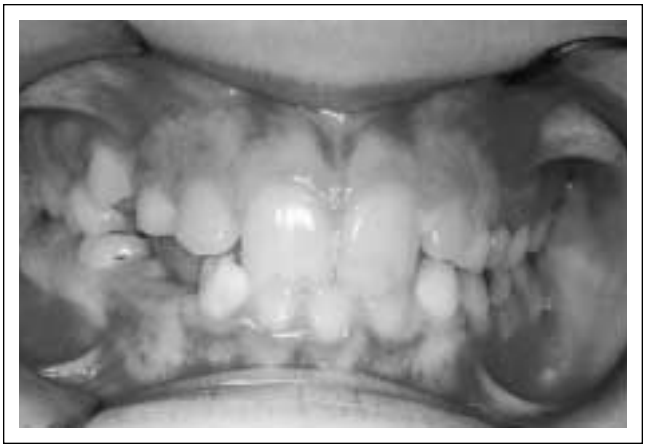
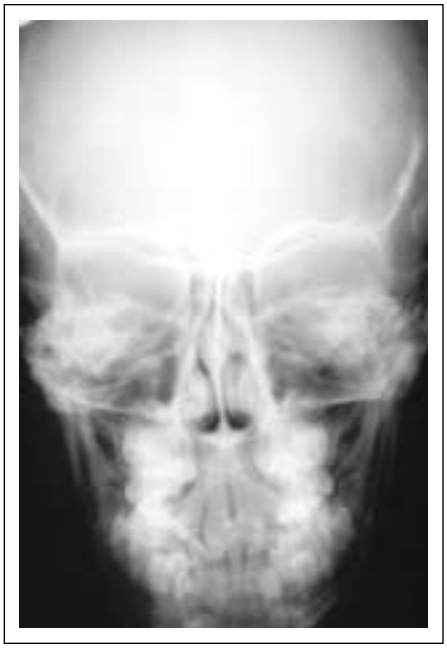


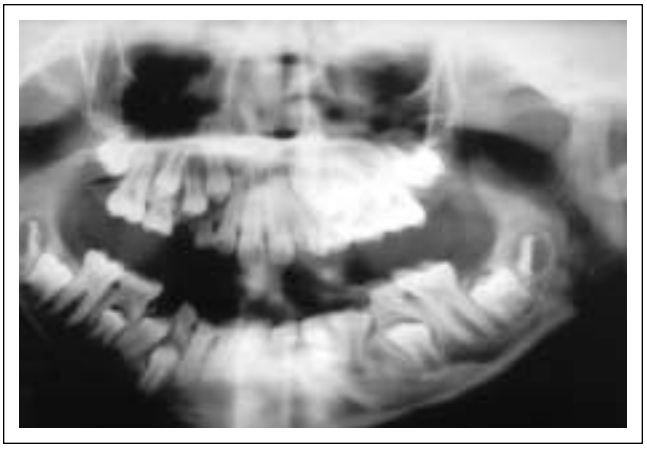
Figure 4. Intraoral view: a mixed-dentition stage with a first molar class and a deviation of 2mm of the mandibular midline in central occlusion are present.

main complaint of the patient was an increasing difficulty in performing normal mandibular movements and an increasing reduction in mouth opening, particularly during the last six months. In the same period occasional pain in the right TMJ during mouth openings was referred, with an increasing frequency in the last three weeks which made it suddenly perceived as a problem.

The clinical examination showed skin lesions on the right cheek (Figure 1), a reduction in mouth opening (up to 30mm) with a deviation of the midline (3mm) and a moderate shift of the chin to the right (Figure 2). Protrusive excursions were normal, while lateral ones were lightly reduced (7mm) particularly on the right. The occlusal plane was flat (Figure 3). The gentle palpation of the area of the right condyle gave rise to mild pain.

The intraoral examination showed a mixed-dentition stage with a first molar class and a deviation of 2mm of the mandibular midline to the right in central occlusion





Figures 5-6. The panoramic radiography and the postero-anterior cephalometric projection confirm a light facial asymmetry.

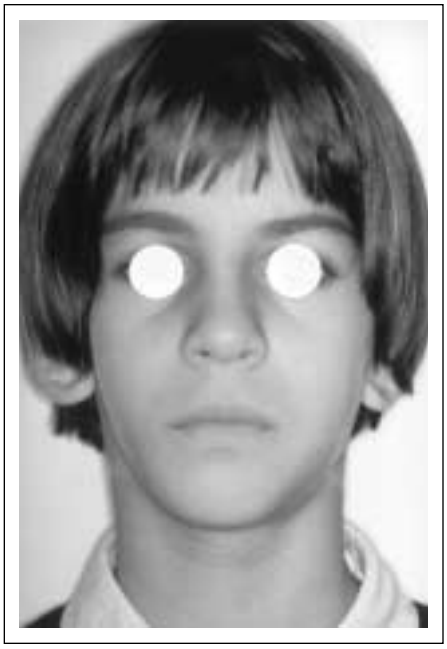


Figure 7. Frontal view eight months after the beginning of functional appliance therapy: no deviation of the midline and no shift of the chin are present anymore.

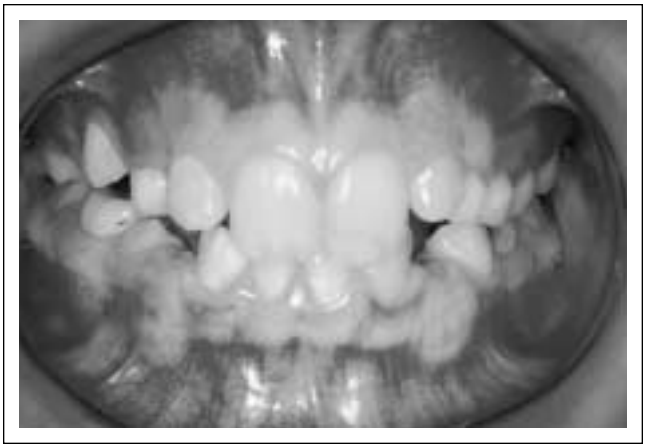


Figure 8. Intraoral view eight months after the beginning of functional appliance therapy: a reduction in mandibular midline deviation in central occlusion is evident.

(Figure 4). The panoramic radiograph and the postero-anterior cephalometric projection confirmed the clinical data (Figures 5, 6).

The girl was immediately scheduled for a functional appliance therapy. Eight months later mouth opening was 37mm, lateral excursions were 8mm on both sides, with no deviation of the midline during mouth movements (Figure 7). No TMJ pain was referred anymore. The intraoral examination showed a reduction in mandibular midline deviation in central occlusion (1mm) (Figure 8).

From this time on, the cooperation of the patient was very poor: she often refused to use the appliance regularly every day, cancelled many appointments and, when recalled, did not come. In the end, the treatment was interrupted.

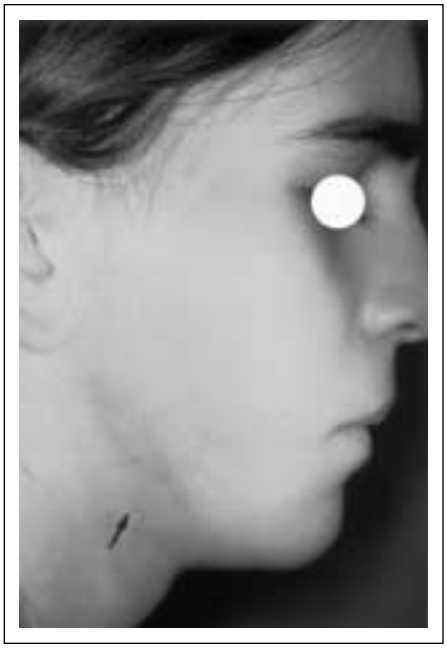


Figure 9. Lateral photograph three years later: the skin lesion is still present.

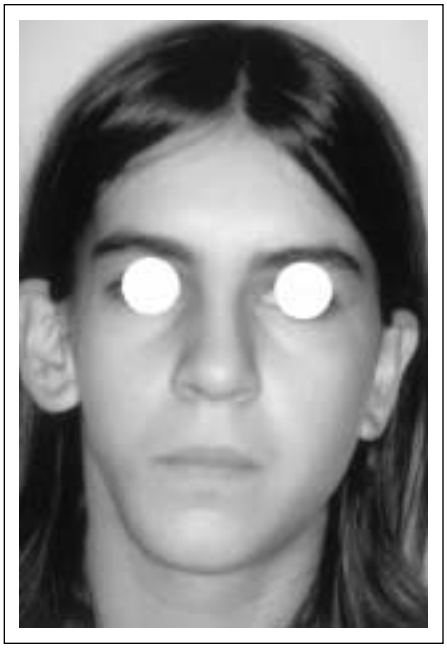


Figure 10. Frontal photograph three years later: a face asymmetry with chin deviation to the right and an atrophy of the masseter muscle are evident.

After three years a new appointment was made because of a developing malocclusion with facial asymmetry. The clinical examination disclosed a facial asymmetry due to a deficiency on the right side, with some initial, apparent effect on adjacent areas of the maxilla. The atrophy of the masticatory muscles on the right side of the face (particularly the masseter) (Figures 9, 10) and a marked misalignment of the occlusal plane with a tip to the right were evident (Figure 11). Mouth opening was 32mm with a shift of the chin to the right. A laterodeviation of 3mm during mouth opening was present; lateral excursions on the right were reduced to 7mm., protrusive ones were normal. The intraoral examination showed a permanent dentition stage with a bilateral first molar class, the ectopic eruption of the 2.3 and a deviation of the mandibular midline in central occlusion of about 2mm. (Figure 12).

The postero-anterior cephalometric projection put in evidence an initial undevelopment of the ramus on the right resulting in moderate decrease in vertical height and mandibular asymmetry (Figure 13).

The patient was scheduled for a fixed orthodontic treatment combined with functional appliance therapy and is still under observation.

DISCUSSION

Any pathology involving the masticatory muscles may result in restriction of mandibular movements with important repercussions on facial development and occlusion. This is due to the intimate interactions existing between bony and soft-tissue structures. In the case described, the use of a functional appliance had excellent results: eight months later, the mouth opening had increased to 37mm and lateral excursions were normal. The occlusal plane was flat, TMJ pain had disappeared and no deviation of the midline during mouth opening was present anymore.

In cases when muscles are the structures to be primarily affected and symptoms are present, functional activation is highly recommended. This treatment provides muscular stimulation to grant continuous function and prevents restriction of articular movements. Of course, it becomes obvious that results are always obtainable if full cooperation is given by the patient.

Unfortunately, in this case, the treatment was interrupted after eight months, as the patient was satisfied with the results obtained and decided not to wear the appliance anymore. During the following three years, she was called for new appointments to monitor the clinical situation, but she never came.

In the end, she decided to start the treatment again because of a developing facial asymmetry combined with malocclusion. At that time, the clinical parameters were remarkably altered: a facial asymmetry due to a deficiency on the right side, with some initial, apparent effect on the adjacent areas of the maxilla was evident; mouth opening had relapsed to 32mm with a shift of the chin to the right; a laterodeviation of 3mm during mouth opening was present and lateral excursions were reduced particularly on the right (7mm). A dysplastic pattern of growth was evident: the occlusal plane was misaligned with a marked tip to the right suggestive of an under development of the ramus on this side and of

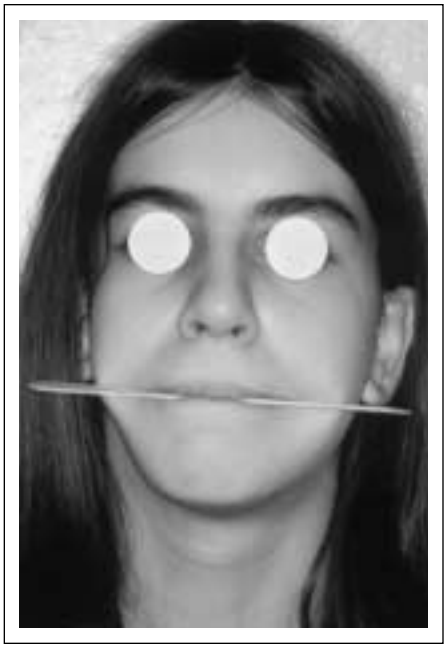


Figure 11. Frontal photograph: a tip of the occlusal plane to the right is evident.

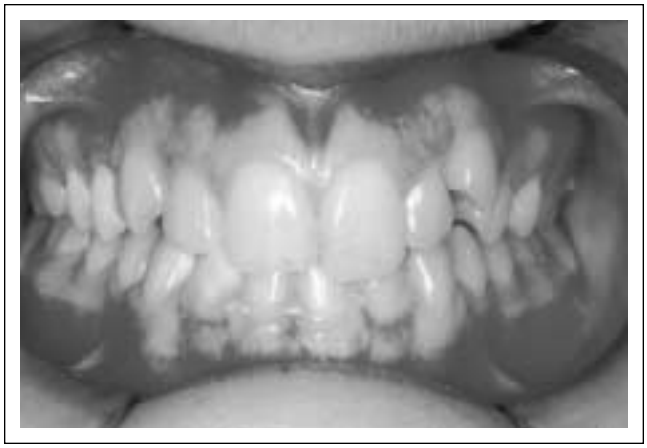


Figure 12. Intraoral view: a permanent dentition stage with a bilateral first molar class, the ectopic eruption of the 2.3 and a deviation of the mandibular midline in central occlusion of about 2mm. are present.

a moderate decrease in vertical height probably due to the atrophy of the masticatory muscles (particularly the masseter).

The intraoral examination showed a permanent dentition stage with a bilateral first molar class, the ectopic eruption of the 2.3 and a deviation of the mandibular midline in central occlusion of about 2mm. The radiographs confirmed the clinical data.

This case clearly shows how the interruption of functional appliance therapy before consolidation of results ends up in a failure. This is probably due to a functional ankylosis of the TMJ related to a muscular dysfunction. An ankylosis like effect on growth is possible even though the mandible is able to move:

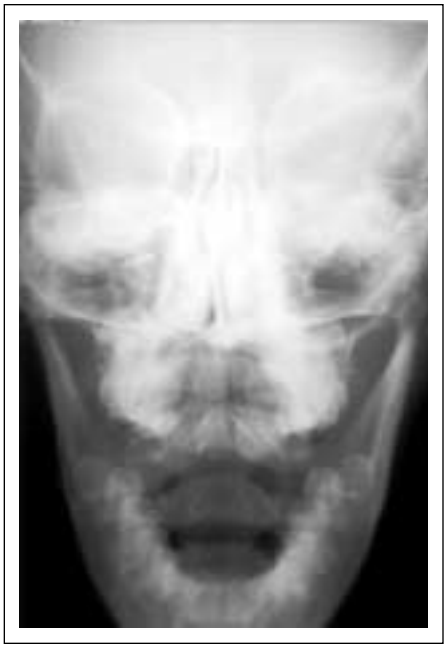


Figure 13. The postero-anterior cephalometric projection put in evidence an initial undevelopment of the ramus on the right resulting in moderate decrease in vertical height and mandibular asymmetry.

a restriction to translate the mandible forward out of the fossa, with functional limitations of the movement, is mainly responsible for that. When opening is restricted to only a hinge type of movement, a progressive growth deficit is often observed and this may result in mandibular deformity and alteration of related structures.

Normal development of the mandible as well as some portions of the upper jaw and face are related to good function of the masticatory apparatus: the integrity and interaction of bony and soft-tissue structures is essential to prevent facial and occlusal disharmonies. From the earliest periods of embryonic growth, an intimate, functional relationship exists between muscles and the bones they are attached. Obviously, as the bones grow, the muscles must change size as adjustment between muscle and bone are to be considered normal part of growth and development. The intimate relationships existing between bones and muscles are mediated through function and require constant adjustment of the attachment between muscle and skeleton.[3] The mandible holds a special interest: there is a general agreement that variations in muscle function markedly affect the areas of muscle attachment with a possible general effect on mandibular size and form. Although the evidence is still not complete, most workers now believe that function plays a more dominant role in the determination of mandibular size and conformation than was previously thought.[4]

All the mandibular muscles involved in jaw movements collectively exert effect on both position and movement of the joint. Posture and movement of the

mandible involve very complex regulation of these muscles; they have two primary functions: contraction and relaxation. Normal muscle function is a matter of contraction or relaxation of individual motor units. Any pathological condition able to interfere with muscular function may result in limitation or inhibition of mandibular movements and end in joint restrictions. Mechanical restrictions and loss of motion in the mandible may result in a loss of stimulus to normal growth: the persistent dysfunction can also lead to disease in the joint secondary to the changes in functional load. When muscular function is altered or impaired the main risk is that of structural changes in the osseous tissue resulting in mandibular pseudoankylosis. These data strongly suggest that the amount of skeletal growth of the face is directly connected to a function equally balanced on both sides and regulated by the functional activation of muscles. For this reason, the mobilisation of tissues within and around the joint is strategic to increase disk mobility and reduce load concentration.[5-6]

In our experience, functional appliance therapy is very useful to correct dysfunction in muscular pattern. It is an excellent, easy way to provide continuous stimulation to the muscles in order to prevent the formation of adhesions within and around the joint and grant a function as much equally balanced on both sides as possible.[7-10] The net forces transferred to the occlusion are dependent on those created by the contraction of the muscles of the mandible. When the plane of occlusion is level,[11] when the mandible, articular disk, and head of the condyles are in good position, and when the neuromuscolar system is in harmony, the loading of forces on the TM joints is optimal and balanced. Disturbances in the harmonious interplay of masticatory muscles may cause a misdirection of the vectors of forces away from the cranial base resulting in unbalanced TMJ loading, sidebending, rotation and torsion of the sphenobasilar synchondrosis.[12] The abnormal transmission of forces to the temporal bones and into the rest of the cranium can be a major factor in creating an externally rotated temporal bone on the low side of the face and internally rotated temporal bone on the high side creating a condition for a progressive hemifacial atrophy. In these cases cosmetic prognosis may be very poor as the dysplastic pattern of growth, if not treated, continues and worsens during the years. The cooperation of the patient is very important to obtain good results: in our opinion, the use of an activator for these patients with the instructions to wear it as many hours a day as possible for at least two years is advisable to correct or minimize effects of muscular dysfunction.

Monitoring of patients during growth is mandatory to intercept further developmental problems.

CONCLUSION

Scleroderma is a rare abnormality in which fibrosis of the skin, subcutaneous tissues and muscle may occur. All forms of scleroderma are rare in childhood: the most common form is localized scleroderma. Localized scleroderma is often benign, but may cause significant deformity if it occurs on the face or extends across joint surfaces. Structural changes may occur in the osseous tissue and result in mandibular joint restriction (pseudoankylosis) and facial and occlusal disharmonies. This article describes a case of facial linear scleroderma in a nine-year-old child and the treatment performed to minimize consequences on occlusion and face development.

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