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Arthrogryposis multiplex congenita (AMC) – case report

Arthrogryposis multiplex congenita (AMC) – is a term describing the presence of various arthrogenic contractures at the moment of birth, limiting, to a certain degree, the range of joints mobility. In some cases several joints can be affected but their mobility might be nearly normal. In classical form of AMC the joints in the following parts of the body become affected: hands, wrists, elbows, arms, hips, feet and knees; in acute form of the disease the joints of the vertebral column become affected which is accompanied by periodical back muscles weakness additionally limiting mobility (1, 2, 4, 8).

In the facial skeleton area there can occur cleft palate, open bite, high-arched palate, limited mobility of the mandible while, generally, no abnormalities in the structure of temporo-mandibular joint are found (6, 7).

The incidence of the disease is low – 1 per 3,000 births. The majority of cases are not genetically determined. The most common form of AMC is amyoplasia, which occurs in 40% of children with arthrogryposis. The real cause of the disease is unknown but, due to reduced functionality of nerve and muscle systems, fetus does not move in uterus which leads to joint contractions in arms and legs (3, 5).

Experiments performed on animals showed that AMC is the result of limited joint mobility during intrauterine life. Lack of normal joint function leads to additional overgrowth of surrounding tissues, which makes the problem even worse. In the situation of joint immobilization the tendons attached to it do not have the possibility of stretching to their normal length and become shorter, which significantly limits joint mobility. Muscle fibers may be replaced by fat or fibrous tissue.

Four groups of causes of the disease are known:

1) disturbances of normal muscle development (atrophy) caused by a congenital disorder, mother's diseases during pregnancy accompanied by high temperature or viral infections destroying cells that transport nervous impulses to muscles;

2) lack of appropriate space for movement during intrauterine life (too little amount of amniotic fluid, abnormal shape of the uterus);

3) deformation of central nervous system and spinal cord (complex form of AMC);

4) abnormal development of tendons, bones, joints, ligaments (incorrect attachment to joint surface)

Case study of a patient diagnosed with AMC

In the year 2000 a 12-year-old patient (K.R.) visited, for the first time, the Paedodontic Dentistry Dept. of the Medical University of Lublin to undergo dental treatment. It was the patient's first visit to a dentist ever. Limited functionality of hands and shoulder joints was found in the patient. His mental development was proceeding normally. The disease had not occurred in any of his relatives.

Dental examination revealed microgenia, limited mobility in temporo-mandibular joints and delayed teething. In the oral cavity there were four permanent first molars, four incisors in the mandible and four in the maxilla, as well as two premolars on the left--hand side of the mandible. The remaining teeth were deciduous canines and molars. Additionally on the molar teeth (bilaterally) occlusion defect of class I according to Angle's classification was observed as well as significant vertical overbite due to excessive tilting of lower incisors. In the upper incisors there occurred centripetal and centrifugal rotations (Fig. 1). DEF mean value of permanent teeth was 4.

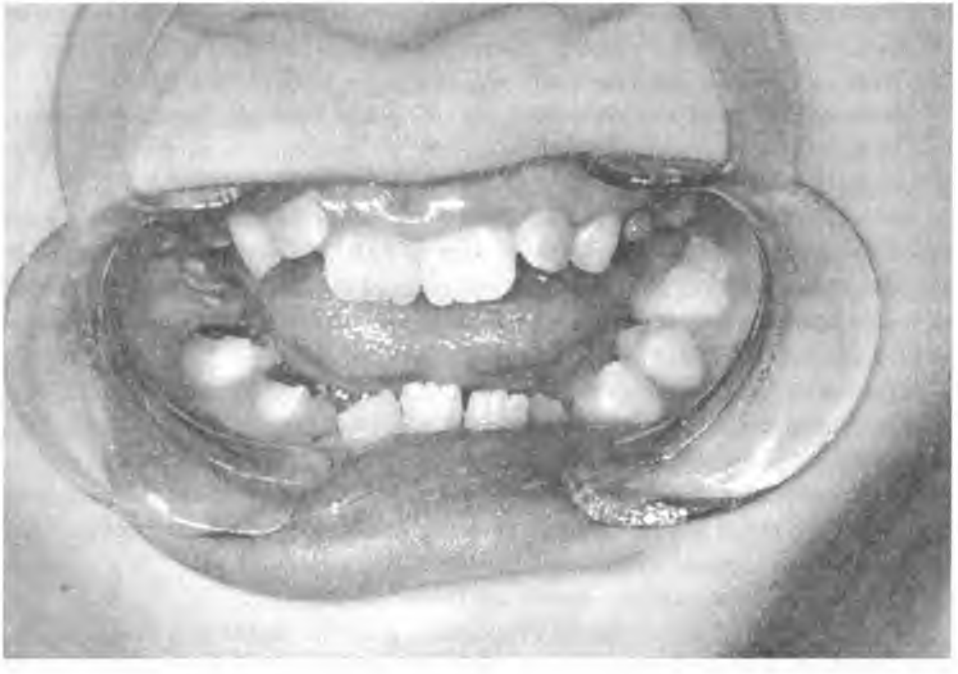

Fig. 1

The conservative and orthodontic treatment applied was difficult due to limited mandible abduction (2–3 cm) and limited lateral movements. In case of other permanent teeth fluoride prophylaxis was used. Wide caries foci were also observed on deciduous molars, which were included in conservative treatment routine. Radiological examination confirmed the presence of buds of the remaining, unerupted teeth (Fig. 2). X-rays of temporo-mandibular joint did not show any significant changes except for the widening of the right occlusal clearance.

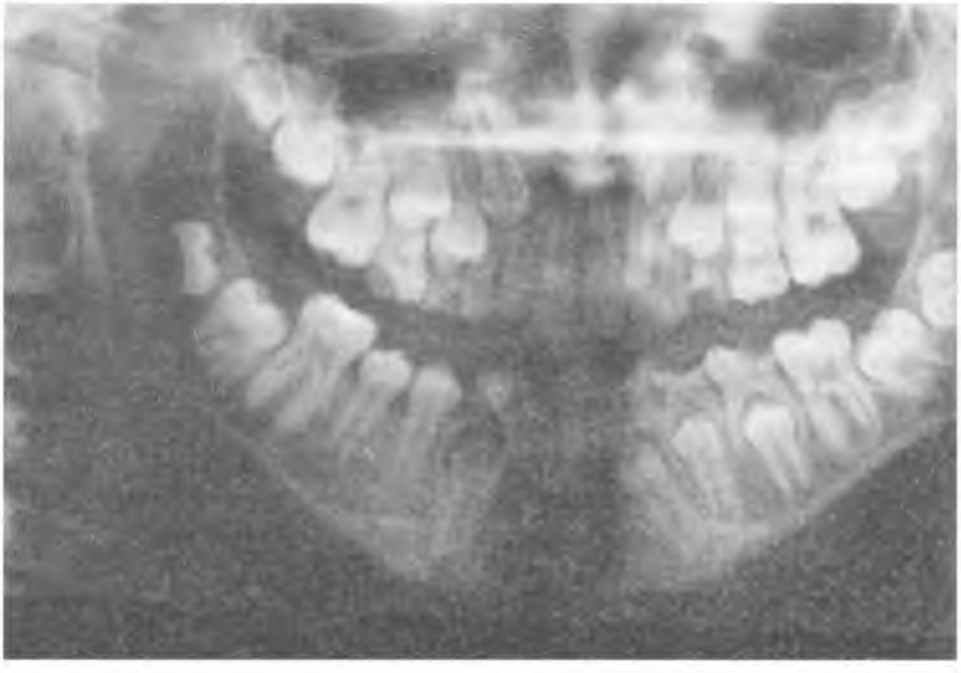

Fig. 2

According to orthodontic indications (lack of space for permanent mandibular canines) deciduous canines, the roots of which had been resorbed by erupting lateral incisors, were extracted. The patient was told to perform muscle exercises to improve the mobility of the mandible (in order to increase the abductive and lateral movements) and orthodontic treatment was implemented.

Children and adolescents with such diseases should remain under constant care of specialists including dentists from the earliest period of their life. Prophylactic activities that are initiated appropriately early can minimize the necessity of very complicated dental treatment required in case of this disease later in life.

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SUMMARY

AMC (which can be also described as fetal muscle cicatrisation) is a term describing the presence of various arthrogenic contractures at the moment of birth, limiting, to a certain degree, the range of joints mobility. The incidence of the disease is low – 1 per 3,000 births and it is usually not determined genetically. In a 12-year-old patient treated in Paedodontic Dentistry Department of the Medical University of Lublin the contractions were observed in elbow joints, wrist joints and ankle joints. Dental examination revealed microgenia, limited mobility in temporo-mandibular joints and delayed teething. DMF mean value of permanent teeth was 4. Earlier the patient has not undergone any dental treatment. The conservative and orthodontic treatment applied was difficult due to limited mandible abduction (2–3 cm) and limited lateral movements. Children and adolescents with such diseases should undergo intensive prophylactic procedures because their dental treatment is very complicated.

Arthrogryposis multiplex congenita (AMC) – opis przypadku

AMC (płodowe zbliznowacenie mięśni) to termin określający obecność różnorodnych przykurczów stawowych stwierdzanych już w momencie urodzenia, ograniczających w różnym stopniu zakres ruchomości stawów. Choroba występuje rzadko – istnieje jeden przypadek na 3000 urodzeń – i zwykle nie jest uwarunkowana genetycznie. U pacjenta 12–letniego leczonego w Zakładzie Stomatologii Wieku Rozwojowego AM w Lublinie przykurcze dotyczyły obu stawów łokciowych, nadgarstków oraz stawów skokowych. Badaniem stomatologicznym stwierdzono mikrogenię, ograniczoną ruchomość w stawach skroniowo–żuchwowych oraz opóźnione ząbkowanie. Liczba PUW w obrębie zębów stałych wynosiła 4. W okresie wcześniejszym pacjent nie był leczony stomatologicznie. Prowadzone leczenie zachowawcze i ortodontyczne było znacznie utrudnione ze względu na ograniczone odwodzenie żuchwy (2–3 cm) i ograniczone ruchy boczne. Pacjentów w wieku rozwojowym, dotkniętych tego typu schorzeniami, należy objąć intensywnymi działaniami profilaktycznymi, gdyż leczenie stomatologiczne jest bardzo skomplikowane.