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Wydawca: Exemplum

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PRACA ORYGINALNA

Kończyna górna i obręcz barkowa

The Hand among criteria of Marfan syndrome and other Marfan-like hereditary elastopathies

Ręka wśród kryteriów zespołu Marfana oraz w innych marfanopodobnych elastopatiach dziedzicznych

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Abstract

Introduction: The Marfan syndrome (MFS) is a genetically determined illness inherited in an autosomal dominant pattern [9,17]. Other elastopathies present in our study are: Ehlers-Danlos syndrome (EDS) – a genetically conditioned disorder of synthesis and metabolism of collagen and other proteins of the connective tissue, Loeys-Dietz syndrome (LDS) – autosomally dominant inherited mutation of the transforming growth factor gene TGFBR1 or TGFBR2, Takayasu Disease (TD) – affects the connective tissue (primarily affects the aorta), congenital ectopia lentis (EL) – autosomally recessive inherited (can also occur in metabolic diseases e.g. homocystinuria) [1-4,5,9,11,12,19].
Aims of the work: 1) Outline of the recognition criteria in frailness syndromes and the need of multidisciplinary care for the patient. 2) Comparison of the hand deformity degree and type between MFS and other Marfan-like syndromes.
Materials and methods: The material comprises of patients under the care of genetics clinics in Poland and the association for patients diagnosed with Marfan syndrome "Marfan Polska" i.e. 144 persons – 72 famale and 72 male at the age between 2 and 70 (77 children, 67 adults). Patients underwent cardiological, ophthalmic, psychological and orthopaedic examination. Apart from anthropometric evaluation and the assessment of joint and vertebral column range of motion the orthopaedic examination included the assessment of the deformities occurring anywhere on the patient's body – function and appearance of the upper limbs included. The study will focus on the assessment of the hand in MFS and on presenting the general criteria of MFS. We would like to emphasize the importance of early interdisciplinary differential diagnosis of MFS and marfan-like syndromes since for a patient who comes to the orthopaedist or the surgeon with signs of arachnodactyly or joint hyper-mobility, early diagnosis can be life-saving. The appearance and function of his hand can be the beginning of the multidisciplinary diagnostics.

Key words: Marfan Syndrome, Ehlers-Danlos Syndrome, Loeys-Dietz Syndrome, elastopatie, joint hipermobility syndrome

Streszczenie

Wstęp: Zespół Marfana (Marfan syndrome – MFS) to genetycznie uwarunkowana choroba dziedziczona autosomalnie dominująco u podstaw, której leży defekt genu FBN1. Do innych elastopatii występujących w naszym materiale należy zespół Ehlersa-Danlosa (EDS) – genetycznie uwarunkowane zaburzenia syntezy i metabolizmu kolagenu oraz innych białek tkanki łącznej; zespół Loeysa-Dietz'a (LDS) – dziedziczony autosomalnie dominująco z mutacja w genie transformującym czynnik wzrostu TGFBR1 albo TGFBR2, choroba Takayashu (ChT) – dotyczy tkanki łącznej (głównie zajmuje aortę), zespół podwichniętej soczewki (ZPS) – dziedziczony autosomalnie recesywnie (może występować w zespołach metabolicznych – homocystynurii).
Cele pracy: 1. Przybliżenie kryteriów rozpoznawania zespołów wiotkości i konieczności wielodyscyplinarnej opieki nad pacjentem. 2. Porównanie stopnia i rodzaju deformacji ręki w MFS oraz w zespołach marfanopodoobnych.
Materiał i metody: Materiał stanowią chorzy objęci opieką poradni genetycznych w Polsce oraz Stowarzyszenia Osób z Zespołem Marfana „Marfan Polska" – 144 chorych, 72 kobiety i 72 mężczyzn w wieku od 2 do 70 lat (dzieci 77, dorośli 67 badanych). Chorzy poddani byli badaniom kardiologicznym, okulistycznym, psychologicznym oraz ortopedycznym. Badanie ortopedyczne oprócz oceny antropometrycznej chorego i oceny ruchomości stawów oraz kręgosłupa, obejmowało ocenę deformacji ortopedycznych całego ciała, w tym funkcji i wyglądu kończyn górnych. Praca ta skupiać będzie się głównie na ocenie ręki w MFS oraz na przybliżeniu ogólnych kryteriów MFS. Chcemy zwrócić w niej uwagę na konieczność wczesnej interdyscyplinarnej diagnostyki MFS i zespołów marfanopodobnych, gdyż wczesne rozpoznanie może być dla chorego trafiającego do ortopedy lub chirurga ręki z powodu arachnodactylii czy hipermobilności stawowej, rozpoznaniem ratującym życie. Wygląd i funkcja ręki może stanowić początek tej wielodyscyplinarnej diagnostyki.

Słowa kluczowe: zespół Marfana, Ehlersa-Danlosa, Loeysa-Dietz'a, elastopatie, zespoły wiotkości

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Introduction

MFS is a genetically determined illness inherited in an autosomal dominant pattern. At its basis lies a defect of the FBN1 gene. It is a systemic damage of tissue (fetal connective tissue) with alterations to the osteoarticular system, cardiovascular system and in the eyeballs. The basis of the illness is a defect in the structure of the connective tissue – fibrilin 1 (a glicoprotein which is the part of elastin fibres). MFS is detected in 1:10000 people. In 75% of diagnosed cases there is a confirmed occurrence in the family which is associated with FBN1 gene on 15th chromosome [5,12,16,17]. According to Polish authors the frequency of MFS is estimated at 1,5-10:100000 persons while the risk of transferring the mutated gene of fibrilin 1 to the offspring amounts to 50% [1,19].

EDS is a group of connective tissue diseases connected with genetically conditioned disorders in synthesis and metabolism of collagen and other proteins included in the connective tissue. There are 8 types of EDS. Type VI – or kyphoskoliotic - is considered marfanoid in fenotype and is associated with a mutation in COL5A1/C gene. The frequency of occurrence of all EDS types is estimated at 1:20000 to 1:100000 births [1,19,20]. When analysing the literature certain sources claim it is significantly more common – 1:5000 and 1:10000 [1,19,20].

LDS has many attributes common with MFS and is caused by a mutation in the transforming growth factor gene type 1 or 2 (TGFBR1, TGFBR2), and is inherited autosomally dominant. Similarly to MFS the EL is connected to a mutation in FBN1 gene but also LTBP2 and ADAMTSI genes. Here Ectopia lentis coexists with skeletal system symptoms but without any cardiological signs (chronic inflamation of large and medium-sized vessels) which dominate in TD; it occurs in patients with HLA-DR2, MB1, HLA-DR4. In Europe occurrence of this illness is estimated at 1-3 cases per million per year. It is considerably more frequent in the countries of Far East – 150 per million per year [9,20].

Modern clinical diagnosis of MFS is based on the Ghent criteria (formulated in Gent, Belhium) which are essential in classifying the patient's illness as Marfan disease or other syndrome with similar symptoms and dysmorphic features [9,12]. They comprise of systemic score in which for each physical feature a number of points are assigned. It is imperative that the patient fulfils the cardiological criteria. MFS can be diagnosed de novo as early as during the interview as long as certain rules are met. In case of negative interview – widening of the aortic trunk (diameter Ao≥2cm) and one of the following: dislocation of the lens with pathogenic mutation of the FBN1 gene and systemic score≥7 points or dislocation of the lens paired with FBN1 gene mutation related to widening of the aorta. When the interview is positive – enlargement of the aorta diameter (Ao≥2cm, age< 20; Ao≥3cm, age≥20), lens dislocation, systemic score≥7 points. If the result of systemic score is lower than 7 and/or aorta diameter has a critical width but no FBN1 gene mutation is present it is suggested to use the term "non-specific connective tissue disorder" until the echocardiographic examination exhibits the enlargement of the aorta diameter. If the mutation is detected but the aorta diameter is not enlarged the term potential Marfan syndrome should be used until the cardiological criterion is met [5,12,19].

Table 1. Ghent Systemic features.

Systemic feature	Points awarded
Wrist AND thumb sign	3
Wrist OR thumb sign	1
Pectus carinatum deformity	2
Pectus excavatum or chest asymmetry	1
Hindfoot deformity	2
Plain pes planus	1
Pneumothorax	2
Dural ectasia	2
Protrusio acetabuli	2
Reduced US/LS AND increased arm/height AND no severe scoliosis	1
Scoliosis or thoracolumbar kyphosis	1
Reduced elbow extension	1
Facial features (3/5) (dolichocephaly, enophthalmos, downslanting palpebral fissures, malar hypoplasia, retrognathia)	1
Skin striae	1
Myopia > 3 diopters	1
Mitral valve prolapse	1

Aims of the work

1) Outline of the recognition criteria in frailness syndromes and the need of multidisciplinary care for the patient.
2) Comparison of the hand deformity degree and type between MFS and other Marfan-like syndromes.

Material and method

The material comprises of patients under the care of genetics clinics in Poland and the association for patients diagnosed with Marfan syndrome "Marfan Polskas" i.e. 144 persons – 72 female, 72 male at the age between 2 and 70 (77 children and 67 adults). Patients underwent cardiological, ophthalmic, psychological and orthopaedic examination. Apart from anthropometric evaluation and estimation of joint and vertebral column range of motion the orthopaedic examination included the assessment of the deformities occurring anywhere on the patient's body – function and appearance

of the upper limbs included – especially the hand Hypermobility of joints was investigated according to the criteria established by Carter and Wilkinson [11]. The evaluation of the hand consisted of appearance assessment, examination of the range of motion (intra-phalyngeal metacarpophalyngeal, carpo-metacarpal joints, passive oposition of the thumb and mobility of carpo-radial joint) [1,5,10,11,12,20]. Patients were divided into two groups – diagnosed with Marfan syndrome and those recognised with marfanoid fenotype: Ehlers-Danlos syndrome (EDS), Loeys-Dietz (LDS), Ectopia lentis, Takayasu Disease (TD) and other non-specific connective tissue disorders (including potential Marfan syndrome (PM)). The former constituted of 71 people (51,07%) diagnosed with MFS. Statistical analysis was performed in Statistica 13 PL computer software.

Results

Of the 144 patients the criteria for MFS were met in 71 cases (40 adults, 31 children). EDS – 10 patients (7,19%), LDS – 2 patients (1,43%), Ectopia lentis – 2 patients (1,43%), TD – 1 patient (1,44%), PM – 54 patients (38,84%) "non-specific connective tissue disorders", healthy – 4 persons (2,15%). In total 79 patients from MFS and marfanoid fenotype groups were diagnosed with disorders of the circulatory system (widening of the aorta in 70 (48,6%) cases, dissection in 9 (6,25%)); vision apparatus disorders – 36 patients (25,01%) from which 33 patients with Ectopia lentis (45,84%). Orthopaedic criteria of MFS were met in 71 (49,3%) patients: 47 (65,29%) exhibited arachnodactyly; among those diagnosed with PM – 31 patients (57,40%). Criteria of joint hipermobility by Carter and Wilkinson [Kirk] wer met by 126 (87,5%) of 144 persons. Patients with arachnodactyly in total – 78 (54%) presented positive wrist-thumb sign. In all those patients the thumb-wrist test gave the result of 180° (Fig.1,2,3). In all MFS and PM patients arm span exceeded their height. The rest of the patients exhibited other disorders of function or appearance of the hand with correct proportions of the limbs. 9 patients (90%) with EDS presented hyperextension in intraphalyngeal and metacarpo-phalyngeal joints ranging to 100° in passive examination and 45° to 60° actively. In 9 patients dorsal carpal extension achieved 120° while palmar flexion was as high as 180° (Fig. 4,5,6). The subjective evaluation of the disorders or function of the limb was also diversified. Patients with MFS and PM did not report pain or sensation of weakness in the hand or the rest of the upper limb. In contrast, all examined patients (children included) with EDS diagnosed reported problems associated with daily manual activity e.g. holding a pen for the duration of 10 minutes. A similar problem with basic functioning in those patients constituted myopia (6 patients ≥6 dioptres). In patients with LDS, deformities of hands and feet concerned shortening of IV and V radius with accompanying camptodactyly; small joints in hands and feet showed narrower range of motion. (Fig. 7,8). In EL 1 patient showed hiperextension in intraphalyngeal and metacarpo-phalyngeal joints (>90°) and radio-carpal joint (100°). In TD no deformations of the hand or function impairment were observed.

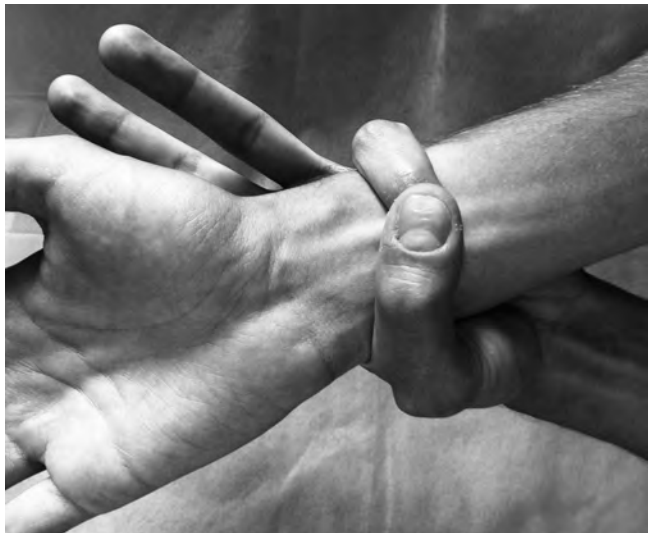

Fig. 1. The hand in MFS.

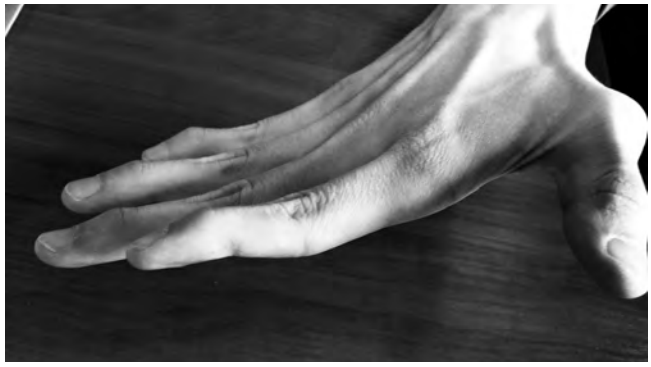

Fig. 2. The hand in MFS.

Fig. 3. The hand in MFS.

Fig. 4. Thumb-wrist test.

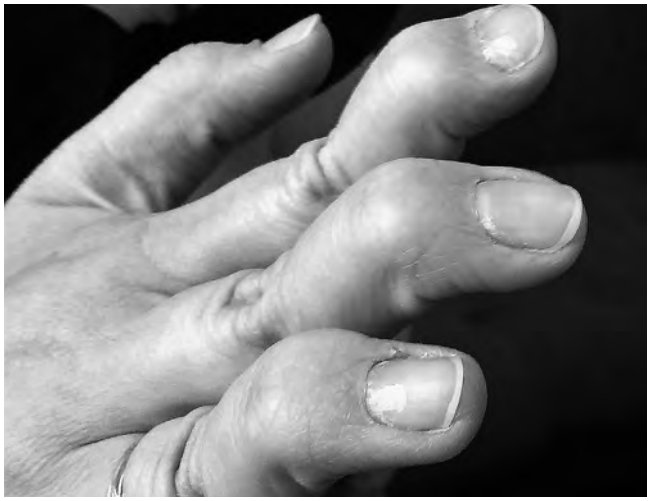

Fig. 5. The hand in EDS.

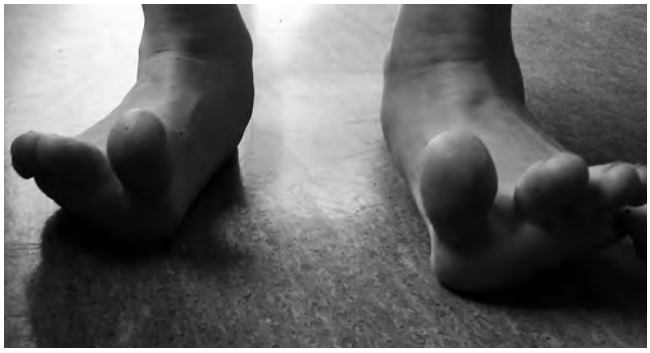

Fig. 6. The foot EDS.

Review

MFS was mentioned in scientific literature for the first time in 1896 when paediatrics professor Antoine Marfan described a case of his five-year-old girl patient named Gabrielle P. She was afflicted with progressive skeletal deformations with a typical appearance of a hand described as congenital contractive arachnodactyly. Over the next decades the clinical symptoms of Gabrielle were supplemented with additional signs observed in similar patients: lens luxation in 1914, congenital nature of the syndrome in 1931, aortic aneurysm and defects of the mitral valve in 1943 etc. until in 1956 McKusick formulated the first widely propagated set of symptoms[12]. In 1986 clinical symptoms and MFS recognition criteria were systematized in Berlin Nosology [12]. In 1996 it was proposed to further improve and tighten them and in 2010 the Ghent criteria were acknowledged as a standard. The diagnostic process of MFS evolved as our knowledge about consecutive organs being affected by the disease improved. It certainly was related to the technical advancement in medicine as far as diagnostic tools are concerned. This phenomenon is called pleiotropy [12]. After more than 100 years since first descriptions of MFS appeared, a number of new organs affected by the disease as well as differential diagnoses became considerably larger. By that we mean syndromes of marfanoid fenotype such as LDS, EL, EDS type VI, TD, MASS fenotype, homocystinuria, Beal syndrome, Achard syndrome, Stickler syndrome and other which weren't mentioned in this study due to them not occuring in our material [1,2,3,5,19,20]. The pleiotropy phenomenon in MFS is evidenced by diagnoses attributed to well-known persons. For example it was said that El Greco, Abraham Lincoln suffered from MFS but today we know his illness was closer to Stickler syndrome [6,13,19], while the case first described by Marfan was in fact Beal syndrome [19]. All of the mentioned syndromes have many common skeletal signs. Taking the hand as an example there is an arachnodactyly in MFS, the so called subjective arachnodactyly (digital shafts and metacarpal bones are narrower but have regular length) in Achard syndrome, arachnodactyly with early signs of arthrosis in intraphalangeal and carpo-metacarpal joints in Stickler syndrome [19], arachnodactyly paired with camptodactyly of IV and V finger with underdeveloped metacarpal bones in LDS [19]. In a group of MFS and PM patients with arachnodactyly that we studied (78 persons (54%)) none reported pain or weakening of the hand function. They also perceived the length of their fingers as a positive feature – just as having a slender figure. Hipermobility of joints and fatigability in EDS were on the other hand viewed as a negative effect of the disease. Arachnodactyly is not present in EDS, TD, homocystinuria, MASS fenotype and FTAAD therefore patients do not report this defect in spite of hipermobility [1-4,13,19]. In MFS with arachnodactyly there is a coexistent hipermobility of interphalangeal, metacarpo-phalangeal, carpo-metacarpal joints and carpus observed [1,5,7-11,14,16,17,18]. A similar situation but with regular length of phalanges and metacarpal bones can be observed in EDS [1,19]. The hand in MFS is often called the Paganini hand – a famous fiddler who owed his success to a huge talent but also the condi-

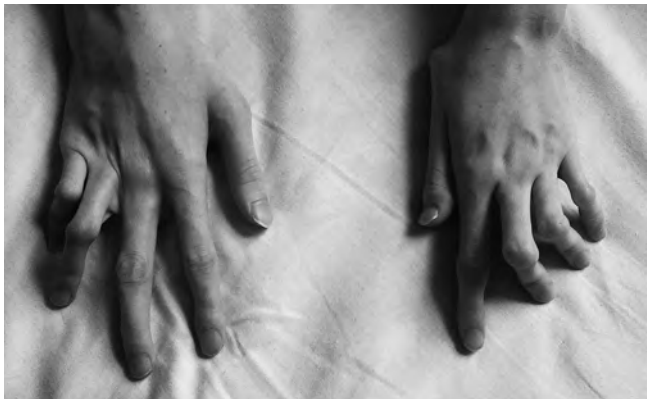

Fig. 7. The hand in LDS.

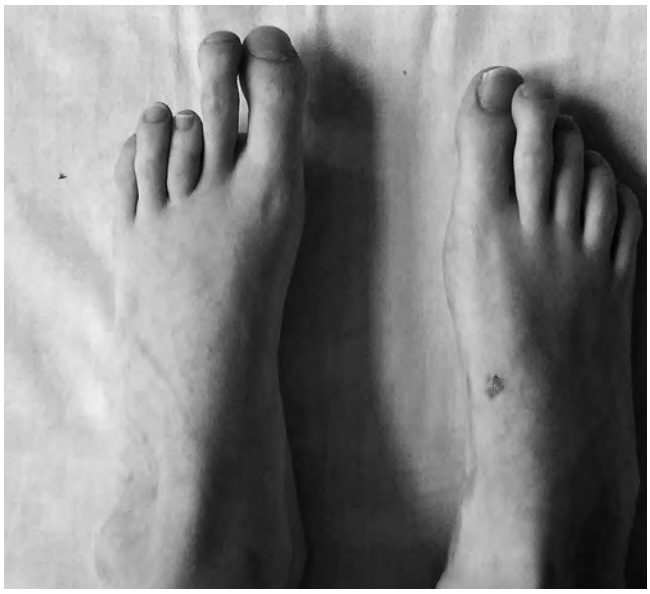

Fig. 8. The foot LDS.

tion of his hands caused by MFS. Paganini's personal doctor wrote: Paganini's hand is not larger than a regular hand but due to its certain parts being more elastic he is able to double his range. For example with\out changing the position of his hand he is able to flex the proximal intraphalangeal joints of the left hand (which touches the string from the side) at the right angle to the natural movement of the joint. He does that without effort, with easiness, confidence and speed (Fig. 9). Essentially Paganini's virtuosity is based on the physical involvement of his hands enriched and developed with never-ending practice [15]. In summary we have to emphasize the importance of early interdisciplinary differential diagnosis of MFS since it can save the patients life. In the material we studied, as of the end of 2016 7 patients were operated because of the aortic aneurysm – it was a treatment considered life-saving. 137 patients from our study require regular cardiological, ophthalmological and orthopaedic check-up with its frequency dependant on the state diagnosed, intensity of symptoms and skeletal deformations.

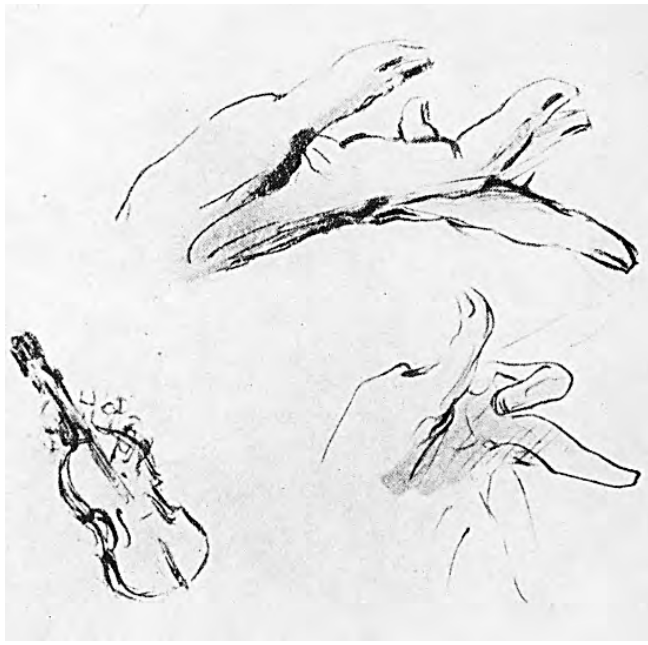

Fig. 9. Paganini's hand [15].

Conclusions

1) The Appearance and function of the hand in congenital elastopathies constitutes a heterogenic group of disorders and plays a vital role in the differentiation of respective syndromes. The appearance of the hand in congenital elastopathies is a a dependable diagnostic cryterion.
2) The type of deformity and functionality disorder in frailness syndromes needs an early diagnostic, individual approach and multidisciplinary care over the patient.

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PRACA POGLĄDOWA

Kończyna górna i obręcz barkowa

Bilateral fracture of radial heads with dislocation treated surgically

Obustronne złamanie głów kości promieniowej z przemieszczeniem leczone operacyjnie

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Abstract

Fracture of the radius is quite frequently observed within elbow joint. This pertains to nearly 20 per cent of acute traumas within elbow joint [2]. Isolated fracture of the radial head occurs in only less than 2 per cent of fractures [3]. Bilateral fracture of radial heads occurs most frequently as a component of severe, high-energy traumas of elbow joint but may also occur after the fall from one's own height. Early diagnosis and minimally invasive surgery allowing for immediate active mobilization of elbow joint are critical in the treatment of radial head fracture. We operated a young female patient with a low-energy trauma, suffering from the fracture of both radial heads, with a method allowing for immediate rehabilitation and obtained a very good functional outcome of treatment.

Key words: fracture, radial head, elbow, displacement

Streszczenie

Złamanie głowy kości promieniowej jest dość często spotykanym złamaniem w obrębie stawu łokciowego. Dotyczy około 20% ostrych obrażeń w obrębie łokcia. Izolowane złamanie głowy kości promieniowej występuje tylko w około 2% złamań. Obustronne złamania głów kości promieniowych występują najczęściej jako składowe ciężkich obrażeń stawu łokciowego po urazach wysokoenergetycznych lecz może wystąpić również po urazie na skutek upadku z własnej wysokości. Najważniejsze w leczeniu złamań głów kości promieniowych jest wczesne rozpoznanie i minimalnie inwazyjna operacja pozwalająca na włączenie wczesnych ruchów w stawie łokciowym. W naszym ośrodku operowaliśmy młodą kobietę po urazie niskoenergetycznym ze złamaniem głów obu kości promieniowych metodą pozwalającą na włączenie wczesnej rehabilitacji uzyskując bardzo dobry funkcjonalny efekt leczenia.

Słowa kluczowe: złamanie, głowa kości promieniowej, łokieć, przemieszczenie

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Introduction

Clinical case

A 42 year-old nurse tripped and fell over on the uneven surface at the operation theatre and sustained an injury of both elbow joints. Radiograms revealed the fracture of both radial heads, assessed according to the Mason's scale type 2.

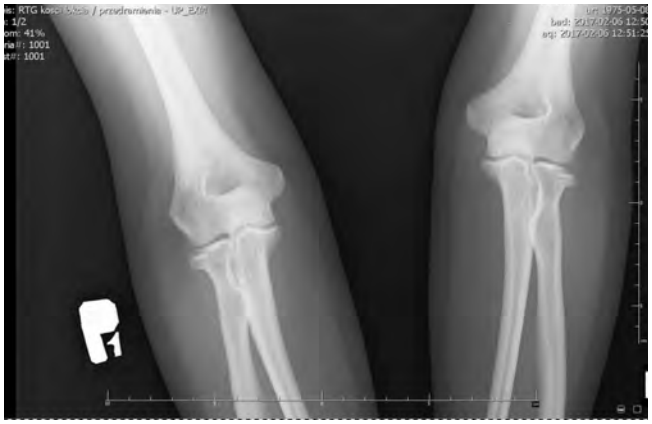

Fig. 1. X-ray picture prior to the surgery.

Patient was qualified for the bloody reposition of fractures and their stabilization by means of metal – compression Medartis screws. Surgery was performed on the fourth day after trauma.

Then – on the second day after the surgery, early kinesiotherapy was applied – active exercises of the elbow joint and wrist along with immobilization in brachio-palmar splints put on for the night.

Ten weeks after trauma radiological characteristics of adhesion were examined and the patient was allowed to do passive and active exercises of the elbow joint and wrist. Four months after trauma an excellent functional outcome of the treatment was reported.

Table 1. Range of mobility of operated extremities

Extremity	Elbow mobility	Supination	Pronation
Upper right	10′-120′	90′	90′
Upper left	5′-130′	90′	90′

Discussion

Bilateral fracture of radial heads is a very rare trauma [8] and usually does not require surgical treatment [9]. Like every fracture of any joint component, Mason II fracture with dislocation requires a surgical intervention [10]. Early kinesiotherapy within the wrist and elbow joints is critical in the treatment of radial head fracture [11,12].

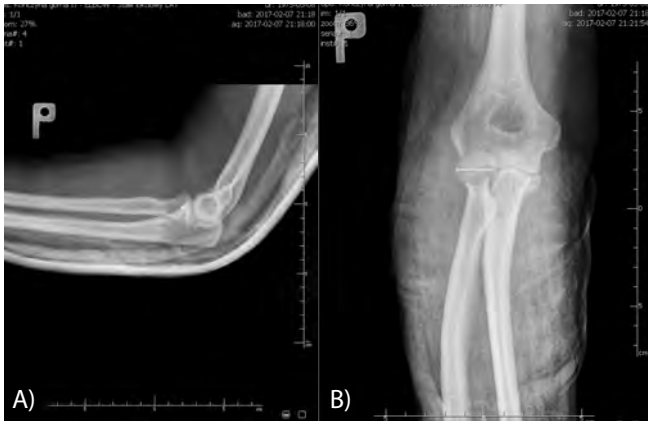

Fig. 2.A-B X-ray picture of right upper extremity after surgery.

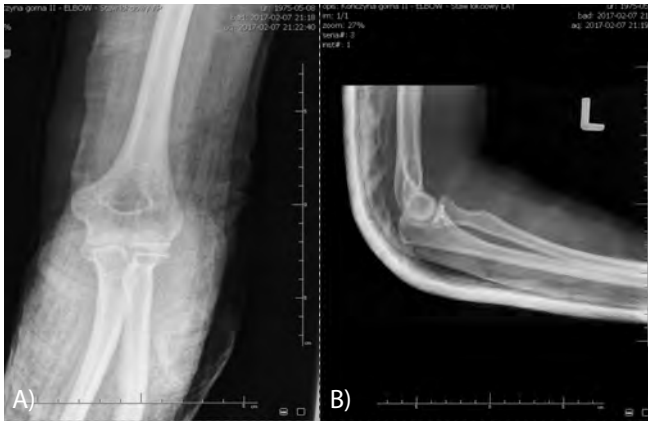

Fig. 3.A-B. X-ray picture of left upper extremity after surgery.

Conclusion

Early radiological diagnostics, low-invasive methods of surgical interventions and immediate kinesiotherapy provide a good functional outcome in patients with Mason II fracture of radial head.

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OPIS PRZYPADKU

Kończyna górna i obręcz barkowa

Paralysis of three upper limb nerves due to a humerus supracondylar fracture – distant results of treatment. Case report

Porażenie trzech nerwów kończyny górnej w przebiegu złamania nadkłyciowego kości ramiennej – odległe wyniki leczenia. Opis przypadku

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Abstract

We are presenting the case of an 8 year old patient who suffered from a left elbow joint trauma and was admitted to the clinic of Children's Orthopedy and Rehabilitation in Lublin due to a left humeral supracondylar fracture in an extensor mechanism injury. Because of this injury, besides damage to the locomotor system, it lead to sensory and motor dysfunction of the three peripheral nerves of upper limb. An emergency closed reduction of the fracture through a percutaneous wire stabilisation was performed. On the third day after the injury, a revision was undertaken of the elbow joint, releasing the trapped brachial artery along with the damaged median nerve. A Kirschner wire was also removed due to conflict with the ulnar nerve. After multiple months of rehabilitation the child regained full left upper limb function.

Key words: supracondylar fracture of the humerus, nerve paresis, vascular dysfunction

Streszczenie

Przedstawiamy opis przebiegu leczenia pacjentki, która w wieku 8 lat doznała urazu stawu łokciowego lewego i została przyjęta do Kliniki Ortopedii i Rehabilitacji Dziecięcej USzD w Lublinie. U chorej doszło do złamania nadkłykciowego kości ramiennej lewej w mechanizmie wyprostnym. W wyniku urazu, oprócz uszkodzenia narządu ruchu, doszło do zaburzenia funkcji czuciowych oraz ruchowych trzech nerwów obwodowych kończyny górnej. W trybie nagłym wykonano repozycję zamkniętą złamania ze przezskórną stabilizacją drutami Kirschnera. W trzeciej dobie od urazu wykonano rewizję stawu łokciowego lewego, uwalniając uwięźniętą tętnicę ramienną wraz z nerwem pośrodkowym oraz usunięto drut K konfliktujący z nerwem łokciowym. Dziecko po wielomiesięcznej rehabilitacji odzyskało pełną funkcję kończyny górnej lewej.

Słowa kluczowe: złamanie nadkłykciowe kości ramiennej, uszkodzenia nerwów, uszkodzenie tętnicy ramiennej

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Introduction

One of the most common fractures in children is the supracondylar fracture of the humeral bone. It constitutes 3% of all fractures and most commonly occurs between the ages of 5-7 [1]. A popular method of evaluating trauma is the Gartland classification, consisting of 3 steps, in which the 3rd degree is defined as a fracture with complete displacement with no cortical contact [1,2]. Treatment of choice in this case would be repositioning of the fracture with internal stabilisation [2]. In case of vascular dysfunction, neural dysfunction or lack of possibility of obtaining the proper alignment, open reposition may be performed [1,2]. Fracture of postero-lateral, postero-medial and rotary displacement with a soft tissue gap are statistically fraught with a higher risk of post operative complications and pre operative nerve dysfunction [1]. Fracture of postero-lateral and rotatory displacement as well as nerve trauma requires physiotherapy and occupational therapy [1].

In 1948 Herbert Seddon divided nerve damage into 3 groups in which the criterium was based on the level of damage of the internal nerve structures. The classification has 3 types of damage: neurapraxia (damage of myelin sheath)- temporary interruption of conduction, which returns to normal after 1-2 days, axonotmesis (axon damage)- blockage of conduction with duration from several weeks to several months usually with full recovery, neurotmesis (endoneurium damage) – regeneration may happen, but isn't always satisfactory [3]. Nervous disorders, commonly neuropraxia, occurs in 3-22% of all humeral supracondylar fractures [2]. A large significance in the damage of particular nerves is due to the mechanism of fracture. In the extensory mechanism there is an increased risk of anterior interosseous nerve neurapraxia, although in the flexor mechanism ulnar nerve neurapraxia is more common. Nerve damage can also be dependant on the way in which stabilisation of bone fragments is performed. Medial percutaneous internal stabilisation of supracondylar humerus fracture can cause an increased risk of ulnar nerve damage[4].

Assumptions and Aims

Discussion of a 2-year long period of observation of the health status of the female patient with post traumatic paralysis of 3 nerves of the upper limb due to humeral supracondylar fracture and presentation of distant treating outcomes.

Materials and Methods

We are introducing the health status of the patient, whom in her eighth year had a trauma to her left elbow joint. The girl has been admitted to the Pediatric Orthopedy and Rehabilitation Department in Lublin due to a left supracondylar humerus fracture in an extensor mechanism injury (Fig. 1).

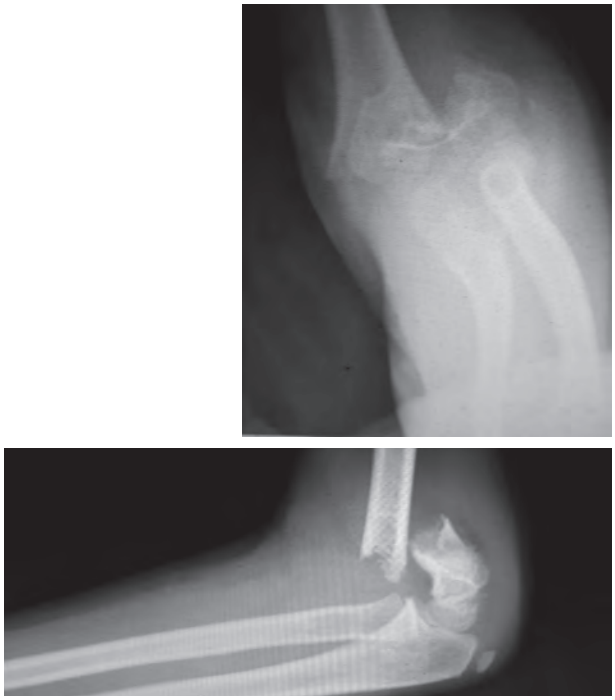

Fig. 1. Supracondylar fracture of the humerus – AP and lateral view.

During clinical examination a large ulnar joint swelling was observed with features of total paresis of median nerve and a partial paresis of the radial nerve. An emergency operation was performed through closed repositioning of the fracture with percutaneous Kirschner wire stabilisation, inserted by crossing the wires from the medial and lateral sides (Fig. 2).

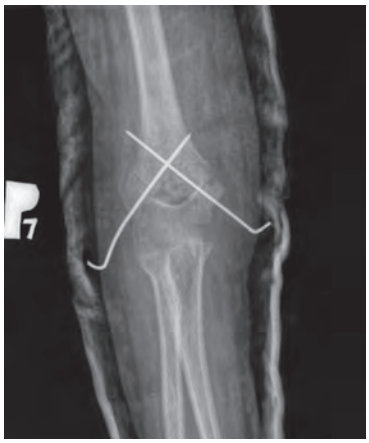

Fig. 2. Closed repositiona of the fracture with percutaneous Kirschner wire stabilisation.

During her third day in hospital a significant decrease in temperature was observed in the circumference of the upper limb and features of total paresis of the median, radial and ulnar nerve. USG Doppler examination showed a decrease in perfusion in the distal part of the limb with no decrease in

saturation of peripheral tissues. The continuity of the nerve was preserved but the conflict with the fixation implants was observed. The radial nerve didn't show a loss of continuity but an edema and obliteration of the fascicular structure of median nerve was visible. Angio-CT examination confirmed features of brachial artery damage. Immediate open revision of elbow joint was performed. During the operation it was determined that the median nerve was damaged, jammed with the brachial artery between the fractured pieces of bone. The brachial artery was released and neurolisis was performed. Neurolysis of the median nerve was carried out and the kirschner wire which was close to the nerve was removed (Fig. 3). Despite obtaining proper positioning of the fractured bone there was no recorded return of all nerve function. During clinical examination it was determined that the active dorsal flexor movement of the wrist was preserved however with loss of extension in the MCP and PIP joints (Fig. 4,5). Radial artery pulse was detectable but the hand was cooler with visible vegetative dysfunction.

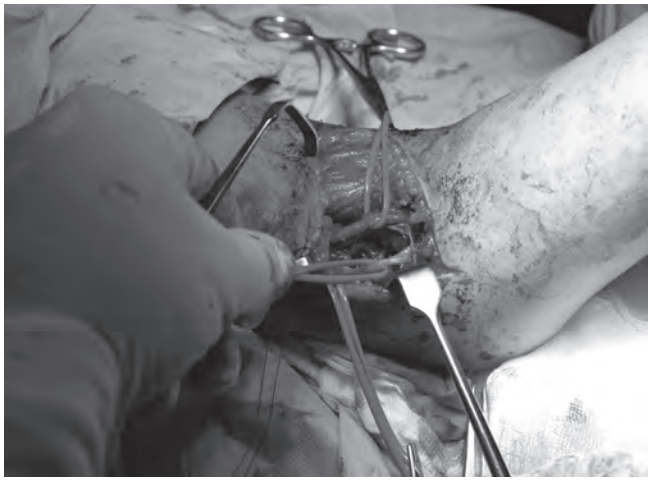

Fig. 3. Revision of cubital fossa – release and nerurolysis of entrapped brachial artery and median nerve.

Fig. 4. Clinical status 3 months after injury.

Fig. 5. Clinical status 6 months after injury

The girl underwent a long lasting, 2 year rehabilitation process. The patient was given multiple galvanising, electrostimulation, hydrotherapy, massage, mobilisation and passive flexor and extensor exercises in DIP, PIP and MCP of the left hand.

Results

Despite bad postoperative prognosis and long lasting and sustained paresis of the median, ulnar and radian nerves there was total remission of symptoms and return to full functionality has been obtained (Fig. 6).

Discussion

Supracondylar humerus fracture is a severe injury of the upper limb fraught with high risk of vascular and nerves dysfunctions. Large displacements should be immediately repositioned, however the final treatment should be performed within 24 hours [2]. Jessica Babal et all ran tests on a group of 5148 people including 5154 of whom had supracondylar humerus fractures. She observed neurapraxia of nerves in 11.3% of patients. Interosseous nerve damage was outweighed in extensor mechanism injury and constituted 34.1% neurapraxia occuring in this mechanism. Ulnar neurapraxia occurred more often in flexor mechanism injury and constituted 91.3% of neurapraxia in this mechanism. Lateral percutaneous internal stabilisation contributed to 3.4% of nerve damage, however 4.1% from the medial approach [4]. Lyons et al. describes a group of 210 children with type 3 supracondylar humerus fractures in extensor injury mechanism among which 19.1% of the patients had either vascular or neurological dysfunctions or both. 13.3% showed nerve damage, 2.9% had nerve damage with coexisting vascular dysfunction and 2.9% had isolated vascular dysfunction.

Fig. 6. Clinical status 24 months after injury – full recovery of radial, median, and ulnar nerve function.

80% of the median nerve damages concerned the anterior interosseous nerve. Damage of the median nerve and vascular dysfunction concerns mainly fractures with postero-lateral dislocation however the damage of the radial nerve coexisted with fractures with postero-medial dislocation [5].

Conclusion

Coexisting damage of radial, median and ulnar nerve with vascular dysfunction occurs extremely rarely and literature does not present unequivocally the frequency of their mutual occurrence. The mechanism of injury and swelling of soft tissues might contribute to nerve paresis [1,5]. The dysfunction of nerves is more often described as neurapraxia rather than axonotmesis [1,4]. Percutaneous internal stabilisation might contribute to iatrogenic nerve damage. An increased number of cases are concerned with damage to the ulnar nerve during insertion of fixation implants from the medial side [4]. With perseverance of continuity of the nerves, long lasting occupational therapy and physical therapy may result in the return of function of the above mentioned nerves.

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PRACA ORYGINALNA

Kończyna dolna i obręcz biodrowa

Bone remodelling in proximal femur after Metha stem implantation – DEXA study

Przebudowa kostna w okolicy bliższego końca kości udowej po implantacji trzpienia przynasadowego METHA – badania DEXA

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Abstract

Introduction: Number of studies showed the high effectiveness of methaphyseal stem implantation in total hip replacement (THR) procedures. This good results inspires us to evaluate the bone behaviour around the methaphyseal stem (METHA Aesculap,Germany) after its implantation.
Aim of study: An evaluation of early bone remodelling process around Metha stem during the first months after THR.
Material and methods: A study group consisted of 28 patients, including 18 women and 10 men at the mean age of 49.6 years, operated for advanced degenerative changes of the hip joint, using the Metha stem implant. The patients were evaluated on the day of surgery, 10 days after and then followed up after 3, and 6 months from surgery by DEXA scanning in seven Gruen zones.
Results: A significant BMD change was found 10 days after the surgery, most pronounced in G3 zone (+37%) and, also, in zones G2 and G5 and the smallest increase in zone G7 zone (3.6%). The first 3 months after surgery showed tendency of BMD reduction in all examined zones. At the period 3 to 6 months after THR a decreasing tendency of the mean BMD values in all the zones, except G6 was observed, while the 6 months DEXA study showed tendency to reverse this reaction.
Conclusions: A short follow-up assessment of periprosthetic bone remodelling after uncemented Metha stem implantation revealed different host-bone responses. The DEXA is a precise method of evaluation of BMD changes around Metha stem.

Key words: Metha stem, DEXA, BMD, bone remodeling, total hip arthroplasty

Streszczenie

Wstęp: Wiele badań wskazuje na dobre wyniki po implantacji trzpieni przynasadowych w aloplastyce stawu biodrowego. Wyniki te zainspirowały nas do oceny zachowania się tkanki kostnej wokół implantowanego trzpienia METHA.
Cel pracy: Celem pracy jest ocena wczesnej przebudowy kostnej w okolicy trzpienia METHA na podstawie badania densytometrycznego.
Materiał i metodyka: Grupa badawcza obejmuje 28 pacjentów, 18 kobiet i 10 mężczyzn, w średnim wieku 49.6 lat w czasie zabiegu operacyjnego. Pacjenci oceniani byli klinicznie, radiologicznie oraz badaniem DEXA po 10 dniach, 3 i 6 miesiącach po implantacji trzpienia METHA.
Wyniki: Istotne zmiany w BMD stwierdzono w 10 dniu po zabiegu, najbardziej nasilone w strefie G3 (+37%), a także w G2 oraz G5 wg Gruena. Najmniejsze zmiany obserwowano w strefie Gruena G7 (+3.6%). Pierwsze 3 miesiące po zabiegu wykazywały tendencję do redukcji BMD we wszystkich badanych strefach. Pomiędzy 3 a 6 miesiącem po aloplastyce stwierdzono zmniejszającą tendencję do zmian w BMD we wszystkich strefach z wyjątkiem G6. Po 6 miesiącu od zabiegu odnotowaliśmy ponowne wzrastanie BMD.
Wnioski: Ocena przebudowy tkanki kostnej wokół trzpienia przynasadowego METHA wykazała zróżnicowaną reakcję kostną. Badanie DEXA okazało się dokładną metodą w ocenie tych zmian.

Słowa kluczowe: trzpień przynasadowy, aloplastyka bezcementowa, gęstość mineralna kości, DEXA

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Introduction

Total hip replacement is a now a standard method for treatment of advanced stages of hip osteoarthritis which improve patient's function and reducing the pain.

Many recent reports indicated promising clinical outcomes with the use of methaphyseal implants what encouraged us to provide a study about the densitometric scanning of the proximal femur after Metha stem implantation.

Aim of study

The goal of the study was to evaluate an early bone remodelling around the Metha stem during.

Materials and methods

The study group consisted of 28 patients, operated for advanced degenerative changes in the hip joint, using the Metha stem (BBraun, Aesculap, Tuttlingen, Germany). The study group included 18 women and 10 men at the age from 21 to 62 years (the mean age: 49.6 years). All of operated patients had the idiopathic osteoarthritis.

The inclusion criteria encompassed Body Mass Index (BMI) scope within 20-30 and the patient's age till 65 years. All the patients were operated by the same surgeon and implanted a modular stem (monoblock or modular neck with 130° head-neck-shaft (CCD) angle and 0° anteversion). In all the cases, an NSC press-fit acetabular component was implanted and 28 or 32 mm ceramic heads were applied with a medium neck in the majority of cases. No intraoperative complication was noted. The average hospital stay was 7 days and the patients had a similar rehabilitation program from the first day after surgery.

The preoperative diagnostics included the bone mineral density (BMD) in seven Gruen zones around the proximal femur.

The study protocol was approved by the Bioethical Commission (Protocol number. RNN/66/11/KE 12 April 2011)

Study methods

In all the patients, qualified to Metha stem implantation, beside standard, preoperative x-ray images, patient's assessment by densitometry (DEXA) was performed of the hip joint, planned for surgery. Then, on the 10th day after surgery, densitometry of the operated hip joint was carried out in the orthopaedic programme only. The patients were then followed by DEXA evaluation after3, and 6 months after the surgery.

Densitometric examination

Proximal femur bone densitometry was performed using a Lunar PRODIGY scanner (GE Medical Systems) with the dedicated enCore software, C 10/2003 version. The scanner was standardised every day, following the manufacturer's recommendations. An examined limb was each time stabilised with a special adapter in 20° internal rotation to ensure the same, repeated limb position in each scanning [1,2]. The first densitometry was carried out on the day of patient's admission as part of a preliminary diagnostic package. A subsequent densitometric scanning was carried out using the orthopaedic program, where the scanner automatically designated 7 Gruen zones around the implanted stem, following an optional adjustment of the long stem axis in the program. Gruen zones were adapted to the short stem design (R1–R7).

The obtained stem images were plotted on the picture, obtained before the surgery, having it first converted to the orthopaedic format. That approach enabled a direct comparison of the dynamics of post-operative changes in particular Gruen zones, also vs. their preoperative baseline BMD values. All the measurements were stored as enCore files and hardcopies.

Statistical evaluation

A statistical analysis as Student's-t distribution, the Wilcoxon test and the Shapiro-Wilk test were used in this study. The level o statistical significance was accepted at α=0.05.

Results

All the qualified patients completed the study according to schedule. No signs were observed of radiographic stem migration or radiolucent lines around the stem. In the course of conducted observations, we found considerable differences in the dynamics of changes around the stem, should the point of reference be the preoperative (baseline) status or the post-operative status (10 days after), that leads to different results in statistical analysis. While analysing the percentage changes in particular time intervals, there was a significant BMD change immediately after the surgery, most pronounced in G3 zone (+34%) and, consecutively, in G2 and G5 zones (30% and 22%, respectively), that could be attributed to the highest compression of the spongy bone in those zones during the stem implantation. The smallest post-operative BMD increase was identified in G7 zone (3.66%).

Densitometry results

Our evaluation concentrated on the mean quantitative results of DEXA measurements in particular Gruen zones, the percentage changes in the time intervals between subsequent controls and the dynamics of the changes in selected observation periods. The dynamics of metabolic processes around the stem was evaluated vs. its status before surgery (see Table 1).

In the course of the study, we found considerable differences in the dynamics of changes around the Metha stem, depending whether the reference point is the preoperative status or the status on the 10th day after the operations, that leads to different results in statistical analysis. Analysing the percentage changes in particular time intervals, one may see a big BMD changes immediately after the surgery, most pronounced in G3 zone (+36%) and, consecutively, in G2 and G5 zones, by 30% and 22%, respectively, that may have been attributed to the highest compression of the spongy bone in those zones during stem implantation. The smallest, post-operative BMD increase was noted in G7 zone (3.66%).

In the first 3 months after the surgery, bone density demonstrates an overt dropping tendency at all the studied zones. That phenomenon of „osteolysis" varied from -1.2% at G6 zone up to -17% at G7 zone. In the second quarter after the surgery, further BMD decrease was observed at all but G6 zones but with lower dynamics and varying from -5.8% at G7 to +0.6 at G6 zone.

The end of the 6 month of the showed a reversal tendency with mean BMD values increasing at particular area of G1-G6 zones. Those values varied from 0.9% at G4 zone to 3.3% at G1 zone. Only G7 demonstrated further BMD decrease by 1.9%.

Discussion

Today we have anhigh number of available THR's models, from standard designs, through hip resurfacing, up to metaphyseal stems which have been gaining more and more advocates in the world of orthopaedic surgery. In all cases, the most important issue is an optimal healing of implanted elements into patient's osseous bed [3,4,5,6].

For this reason, we decided to evaluate early bone remodelling process around the Metha stem during a 6-month period after its implantation.

In our study, early periprosthetic bone behaviour around the Metha stem was observed and its relationship with the preoperative bone mineral density (BMD) was investigated in the femoral metaphyseal area. The DEXA is considered the most reliable tool to evaluate bone remodelling after THA with different stem designs [7].

During the recent years, a number of reports have been published with evaluation of bone remodelling processes at stem implantation areas. Lerch et al. evaluated bone density around BiCONTACT stems, Albanese et al. analysed CFP, Mayo and IPS stems, while Brinkman et al. made Nanos stems their issue of interest [4,5,6].

The analysis of Gruen zones is the most commonly used method to evaluate bone remodelling or aseptic loosening after implantation of femoral stems [7]. According to literature reports, conventional Gruen zones were adapted to the short stem design [8,9].

The available literature offers reports, presenting bone remodelling around implanted stem in long-term follow up after operation [10,11,12]. In our opinion, the observation of bone tissue remodelling around the implanted stem is most important when performed in the early, post-operative period, therefore, similarly as Mulier, we evaluated bone remodelling around the Metha stem during the period of 10 days, as well as during 3, 6 and 12 months after surgery [13].

In our study, no other factors were analysed, which may have had any influence on bone tissue remodelling around the Metha stem. It does appear from the available literature, whether such factors, as sex, body weight or the type of implanted stem, play any significant role in bone tissue remodelling around the Metha stem [14, 15,16].

Another important issue in our studies was the reference of BMD changes around the stem to their preoperative (baseline) values and to their status after 10 days from the operation. The changes in bone density, which occur in the metaphyseal part of the femoral bone, are associated with the surgical technique of site compacting to implant the Metha stem. Similar observations were made by Leichtle, although he claims from results from experimental studies that compacting of trabecular bone or bone loss, due to rasping are not the main causes of density changes, while blood-flow disorders, following femoral preparation, may affect some

Table 1. Mean quantitative results of DEXA measurements in particular Gruen zones (g/cm^2).

	G1	G2	G3	G4	G5	G6	G7
Before	0.79184	1.00292	1.56712	1.7966	1.55144	1.24824	1.18652
10 day	0.860964	1.303643	2.126464	2.044464	1.891036	1.497929	1.229821
3 months	0.783519	1.219963	2.023852	1.993963	1.824222	1.479667	1.021333
6 months	0.746103	1.181483	2.008172	1.93031	1.778207	1.488207	0.961931

of Gruen zones, causing bone density drop during the first months after surgery [17].

We believe that, clinical values may be attributed to considerations on the dynamics of metabolic processes vs. their post-operative status. It is so, since after operation, there is an „artificial" increase of bone density in particular zones, obtained the surgical technique itself (compacting the spongy bone to obtain an appropriate site in the femur bone for stem implantation). Further deliberations have been based on that premise, namely that the „independent" bone metabolism around the stem should be evaluated in observations of the status immediately after operation (10 days). Nevertheless, an interesting information can be found in the data, comparing the mean bone density in G1-G7 zones after 10 days from the operation vs. its preoperative values. The data give us a picture of BMD changes in these areas, induced by the operative technique, where the highest post-operative BMD increase can be seen in the lateral (G2 and G3) and medial (G5 and G6) zones [19,20].

Our observations demonstrated a slow BMD drop in all the Gruen zones during the first three months from the surgery. After 4 months, the BMD decreasing trend continued in the observed zones, however, with a slower pace. From the 6th month after the Metha stem implantation on, a new tendency was observed towards a gradual BMD increase in the observed zones.

In the studies, presented by Zah et al. with the use of a Nanoss stem it appears that DEXA scan showed a significant and relevant increase in G-6 zone (12%) and a decrease in G-1 (15%), G-2 (5%) and G-7 (12%) zone, which was interpreted as reflecting a distal load transfer in the metaphysis of the femur [4].

Similar observations were made by Albanese et al. In their evaluation of CFP, IPS, and ABG stems, they showed decreased BMD in G-1 zone and, in case of the Mayo, IPS and Alloclassic stems - in G-7 zone [3]. Stukenborg-Colsman et al. also confirmed the biggest BMD drops in the greater trochanter (-11%) and the calcar region (-12%) [19].

In the studies of Lerch, the overall bone mass loss was 2.8% in the entire femur. Bone mass decrease was mostly found in the proximal part of the calcar and in the greater trochanter, probably leading to stress shielding [20].

Pitto et al. found a 39.6% cortical bone mass decrease in the calcar and trochanteric region five years after implantation of a tapered uncemented stem in a qCT investigation and Albanese et al., observed BMD loss in G-7 zone as a known issue, because of stress shielding in the very proximal portion of the femur [3,12,21].

Our study confirms the results obtained by others authors, evaluating BMD levels around the femoral stem [4,12,19,21,22,23]. Our study also confirmed that the DEXA method is a reliable procedure to determine periprosthetic mineral density and may become a useful tool to study bone response to particular THA options.

In conclusion, the assessment of periprosthetic bone remodelling after uncemented Metha stem implantation, obtained in a short follow-up, revealed a different host-bone response. DEXA is a precise evaluation technique, ideal to assess small BMD changes around the Metha stem. The data further highlight the crucial role of mechanical stress in BMD maintenance.

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PRACA POGLĄDOWA

Kończyna dolna i obręcz biodrowa

Symptomatology and diagnosis. Patellofemoral pain

Symptomatologia i diagnostyka. Ból w stawie rzepkowo-udowym

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Abstract

Patellofemoral joint (PFJ) and it's coexisting disorders are an essential clinical problem for orthopaedic surgeons. Literature is lacking of general population epidemiological studies that could confirm an incidence rate of patellofemoral pain syndrome. Available studies are most often taken on a precise group of sport participants. In literature it is written that complaints coming from an anterior aspect of the knee joint are reported by 15-25% of patients connected with sports activity. In the United States of America from 2007 to 2011 7.3 % of all the patients that came forward to medical care were diagnosed with patellofemoral pain syndrome, which means almost 2,2 mln of patients. The most numerous group is women between 50-59 years of age. In addition, between 2007-2011, in the United States of America ,a constant increase of incidence rate was reported in all the population examined in terms of ageing. Anterior knee pain most often affects women, athletes, recreational sport participants (cross-fit, squatting, jogging). The aim of this paper is to sort terminology, symptomatology and rules of differential diagnosis of patellofemoral pain syndrome.

Key words: AKP – anterior knee pain, PFJ – patellofemoral joint, PFS – patellofemoral syndrome, PFP – patellofemoral pain

Streszczenie

Staw rzepkowo-udowy i związane z nim stany chorobowe stanowią istotny problem kliniczny Ortopedów. Istnieje niewiele populacyjnych prac dokumentujących współczynnik chorobowości bólu w przedziale przednim stawu kolanowego, a powstałe doniesienia dotyczą najczęściej określonych grup sportowców. W piśmiennictwie szacuje się, że dolegliwości ze strony stawu rzepkowo-udowego podaje 15-25% osób związanych z uprawianiem sportu [1]. W Stanach Zjednoczonych w latach 2007-2011, u 7,3% chorych zgłaszających się do systemu opieki zdrowotnej rozpoznano PFS (Patellofemoral Pain Syndrome), co stanowi niespełna 2,2 mln pacjentów, wśród których najliczniejszą grupę stanowiły kobiety między 50-59 rokiem życia [2]. Ponadto w latach 2007-2011 w Stanach Zjednoczonych odnotowano stały wzrost liczby chorych, ze współczynnikiem chorobowości rosnącym w poszczególnych grupach wiekowych w miarę starzenia [2]. Ból w przedziale przednim stawu kolanowego dotyczy najczęściej kobiet, wyczynowych sportowców i osób rekreacyjnie uprawiających sport (cross-fit, squatting, jogging) [3]. Celem niniejszego opracowania jest uporządkowanie terminologii, symptomatologii oraz zasad diagnostyki klinicznej bólu w przedziale przednim stawu kolanowego.

Słowa kluczowe: AKP – ból w przedziale przednim staw kolanowego, PFJ – staw rzepkowo-udowy, PFS – syndrom stawu rzepkowo-udowego, PFP – ból w stawie rzepkowo-udowym

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Introduction

Patellofemoral joint (PFJ) and it's coexisting disorders are an essential clinical problem for orthopaedic surgeons. Literature is lacking of general population epidemiological studies that could confirm an incidence rate of patellofemoral pain syndrome. Available studies are most often taken on a precise group of sport participants. In literature it is written that complaints coming from an anterior aspect of the knee joint are reported by 15-25% of patients connected with sports activity [1] . In the United States of America from 2007 to 2011 7,3 % of all the patients that came forward to medical care were diagnosed with patellofemoral pain syndrome, which means almost 2,2 mln of patients [2]. The most numerous group is women between 50-59 years of age [2]. In addition, between 2007-2011, in the United States of America ,a constant increase of incidence rate was reported in all the population examined in terms of ageing [2]. Anterior knee pain most often affects women, athletes, recreational sport participants (cross-fit, squatting, jogging) [3]. The aim of this paper is to sort terminology, symptomatology and rules of differential diagnosis of patellofemoral pain syndrome.

Knowledge

From the local point of view we can focus on anterior knee pain – AKP, but pathology of the patellofemoral joint outreaches wider than only the joint itself [4]. In this connection patellofemoral syndrome – PFS – should be defined, which refers not only to the symptoms coming from the knee, but also those that are caused by other dysfunctions – hip joint, foot, lumbosacral part of the spine [4,5,6,7]. AKP the local point of view, PFS the global one, are commonly used interchangeably in the literature, which in essence and complexity of pathophysiology of PFS brings some confusion and also suggests limitation to local pathologies of the knee joint. This can cause diagnostic and therapeutic mistakes.

In 2016 The Patellofemoral Pain Consensus Statement from the 4th International Patellofemoral Pain Research Retreat, Manchester was published. It qualifies definitions:

1. Preferred term for denomination of the pain localized in the anterior aspect of the knee is Patellofemoral Pain – PFP. It is a synonym of following terms: AKP, Patellofemoral Pain Syndrome, Chondromalacia Patellae, Runner's Knee.
2. The core criterion required to define PFP is pain around or behind the patella, which is aggravated by at least one activity that loads the patellofemoral joint during weight bearing on a flexed knee (eg, squatting, stair ambulation, jogging/running, hopping/jumping)
3. Additional criteria – not essential to make diagnosis – crepitus or grinding sensation emanating from the patellofemoral joint during knee flexion movements, tenderness on patellar facet palpation, pain on sitting, rising on sitting, or straightening the knee after sitting [8].

To consider PFJ it is mandatory to know the exact anatomy and biomechanics of the joint. Patella is the biggest sesamoid bone in the human body which composes a patellofemoral joint with the patellar articular surface of the femur that creates trochlea for movements of the patella – intertrochlear groove [9]. Trochlear angle is individually variable, normal bony angle is 138 +/- 6 degrees [6]. Bony trochlear angle may differ from the cartilaginous one [9]. In distal direction trochlea deepens. Lateral femoral condyle is bigger than the medial one [6]. Medial Patellofemoral Ligament (MPFL) is in charge of prevention of lateral dislocation of the patella to the knee bent to an angle of 20 degrees and assures 60% stability [10]. Another 20 % is provided by medial patellomeniscal ligament [10]. The more the knee is bent the more bony stabilization patella gets from the intercondylar groove [6,9]. Patellar traction in PFJ is controlled by active muscle contraction: Vastus Medialis Obliquus (VMO), which is the biggest active stabilizer in PFJ. It also gives branches to the MPFL, thus it takes part in proprioception, Rectus Femoris (RF), Vastus Lateralis (VL). Passive stabilization comes from Patellar Tendon (PT), Iliotibial Band (ITB), lateral retinaculum, MPFL, bony stabilization [6,9,11]. Patellar articular surface changes the contact area from the distal part of the patella to its proximal aspect according to the knee flexion angle [9]. In PFJ patella is a lever arm for the knee extensor mechanism, which transmits high loads with force adequate to support seven times body weight [9,12]. It corresponds to cartilage thickness in PFJ, which is 4-5 mm [10].

Differential diagnosis of PFJ disorders consist of patient interview and physical examination [13]. Essential tips from the interview are: character of complaints, duration, factors to aggravate and relief symptoms, traumatic history especially direct anterior trauma, sprain, acute or chronic patellar instability [14]. Patients often declare theatre sign - AKP after long periods of sitting with the knee bent at a 90 degree angle (long journeys, work positions), pain during stair climbing or descending, sensation of instability or giving-way [12,14]. In sport related injuries key information is concerning changing training habits, frequency and also equipment modifications, especially running shoes. Athletes competing in jumping activities such as basketball. volleyball, handball, football are vulnerable [11].

Physical examination must be taken according to specific arrangement which guarantees its thoroughness. It starts with inspecting the patient standing form the anterior and posterior aspect. We can evaluate the length of the lower limbs, its rotation and alignment, pelvis placing, foot adjustment (e.g. overpronation, pes plano-valgus), adduction in the hip joint. These are factors that cause an increase of an Q-angle, lateral patellar compression and as a consequence maltracking in

PFJ [15]. Next step is evaluating gait habits and its correctness [14]. Examination should be continued in a sitting position checking both patellar placement its disparity and height. In dynamic tests we search for PFJ instability symptoms such as 'J-sign' – lateral patellar subluxation during knee extension which indicates insufficiency of medial structures. We measure Q angle (M: 14 degrees, W: 18 degrees) [15,16]. During palpation of the patella and forcing compression in PFJ (Patellar Grind Test, Clarke Test) in dynamic examination it is possible to feel the crepitus protruding from under the patella, which are most often caused by patellar chondromalacia, plica syndrome [17]. Other tests are lateral and medial glide tests, examined in extension and 30 degrees of flexion [15]. Excessive medial subluxation tells of lateral retinaculum insufficiency, excessive lateral subluxation is an evidence of medial stabilizers insufficiency – MPFL. Exaggerated reduction of range of motion can be caused by arthrofibrosis or lateral retinaculum fibrosis. In this connection passive patellar tilt test should be performed. We try to elevate the lateral ridge of the patella, trying to rotate it internally [18]. Checking the way that patella enters the intercondylar groove as the knee deepens flexion may reveal clicking, crepitus or the patient complains of pain that suggests trochlear dysplasia (TD). An 'apprehension test' is performed by trying to luxate the patella laterally. Patient reacts with fear of luxation and contracts extensor muscles not allowing the continuation of the test [15]. In addition to the manipulation mentioned above it is mandatory to perform complex knee examination with meniscal tests, medial , lateral and central stabilizers function. In the literature the most effective test in diagnosis of the PFS is a 'squatting test', performed by asking the patient to do a deep squat with the examined leg positioned forward with the knee flexed to 90 degrees [8]. Pain caused by the test is present in about 80 % of patients with PFP. Significance of all the other tests is limited [8,16,17].

Properly taken clinical examination should comprise of a diagnosis. Additional studies and imaging tests are taken for confirmation or to hammer out a solution.

A basic diagnostic step in PFJ evaluation is an x-ray. First of all, basic anteroposterior and lateral views should be performed. Furthermore Merchant (sunrise view) view should also be performed [10]. To make a certain diagnosis, it is mandatory to take x-rays in correct positions according to general known rules. Especially it refers to a lateral x-ray of the knee joint, which discloses the posterior aspect of the femoral condyles as one (true lateral view). Mentioned x-rays provide essential information to establish basic parameters [10]. The least information comes from an anteroposterior x-ray. Evaluation of severe pathologies is possible: high grade patella alta or baja, patella bipartite [10]. On the true lateral view precise measuring of parameters evaluating patellar positioning is available: Insall-Salvati (IS) (patellar height and patellar tendon index) and Canton-Deschamps (CD) (articular surface of the patella to a distance from the apex to tibial plateau) for diagnosis of patella baja <0,8 IS, < 0,6 CD, patella alta >1,2 IS, >1,3 CD [10,19] (Fig. 1,2). Furthermore a true lateral view offers an evaluation of correctness of an anatomical features of the knee joint or reveals TD, according to Dejour from type A to D [9] (Fig 3).

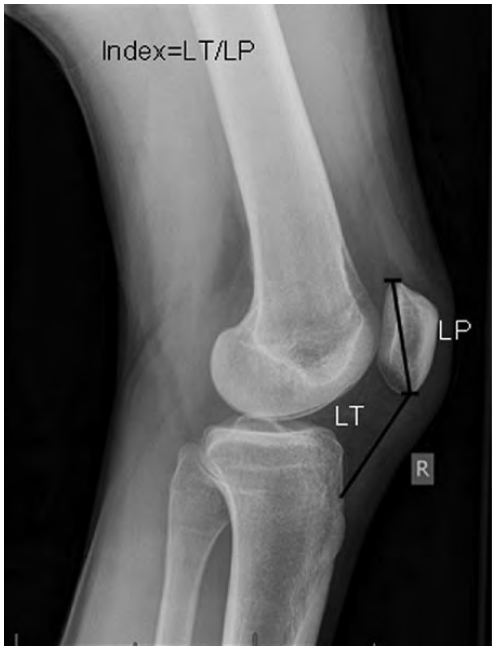

Fig. 1. Insall-Salvati Index, Zaffagnini S, Dejour D, Arendt E, Patellofemoral Pain Instability, and Arthritis Clinical Presentation, Imaging, and Treatment, Springer-Verlag Berlin Heidelberg 2010, page 62, figure. 7.1, used with kind permission of the Editors.

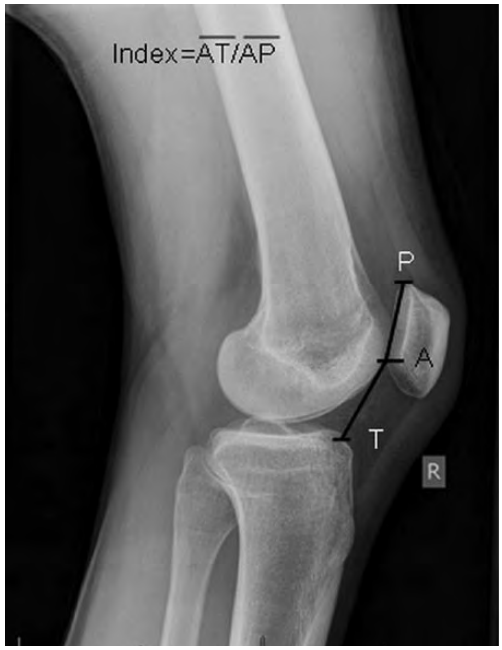

Fig. 2. Canton-Deschamps Index, Zaffagnini S, Dejour D, Arendt E, Patellofemoral Pain Instability, and Arthritis Clinical Presentation, Imaging, and Treatment, Springer-Verlag Berlin Heidelberg 2010, page 63, figure 7.3, used with kind permission of the Editors.

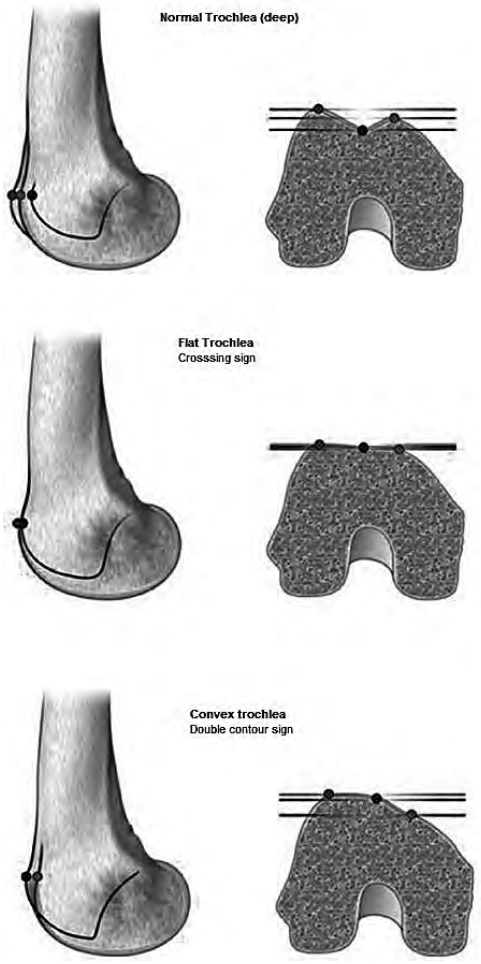

Fig. 3. The crossing sign defines the trochlear dysplasia: the groove line links the condyles anterior part; at that point the trochlea is flat or convex, Zaffagnini S, Dejour D, Arendt E, Patellofemoral Pain Instability, and Arthritis Clinical Presentation, Imaging, and Treatment, Springer-Verlag Berlin Heidelberg 2010, page 53, figure 6.3, used with kind permission of the Editors.

In type A, on a true lateral view a crossing sign is seen. It tells of a shallow intercondylar groove, type B with supratrochlear spur that is a sign of a flat trochlea, type C which is made of a combination of crossing sign and double contour sign. It is made by hypoplastic medial femoral condyle. Type D that ties up all the features mentioned – crossing sign, supratrochlear spur and double contour – as an expression of complete TD [9] (Fig. 4).

Sunrise view of Merchant shows: the type of patella according to Wiberg classification, osteoarthritic changes in PFJ, patellar tilt [9]. Performing Merchant view with 20 degrees of flexion instead of 30 , before the patella gets bony stabilization in trochlea reveals features of patellar instability (MPFL insufficiency).

An additional x-ray in PFJ pathology is standing, weight-bearing anteroposterior hip to ankle view, known as HKA (hip, knee, ankle), allowing to draw Maquet's line. Axial varus malalignment or especially valgus cause maltraction in

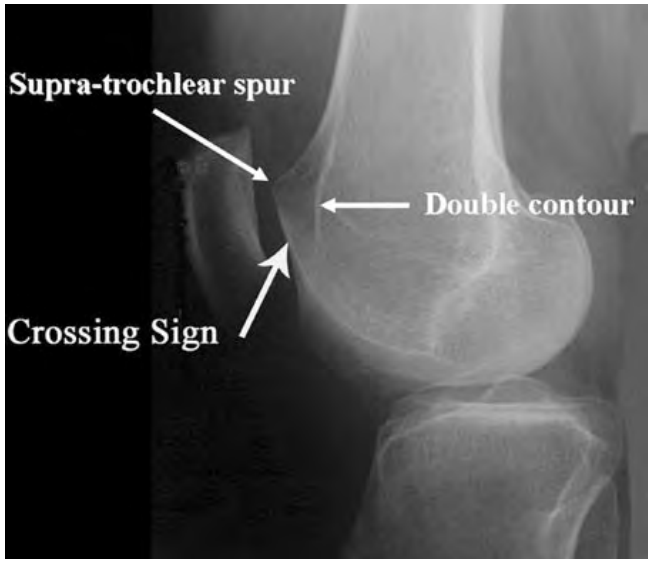

Fig. 4. The three patterns of trochlear dysplasia: Crossing sign; Supratrochlear spur: defines the prominence of the trochlea; Double contour: projection on the lateral view of the hypoplastic medial facet (subchondral bone); it should be below the crossing to be pathological, Zaffagnini S, Dejour D, Arendt E, Patellofemoral Pain Instability, and Arthritis Clinical Presentation, Imaging, and Treatment, Springer-Verlag Berlin Heidelberg 2010, page 54, figure 5.4, used with kind permission of the Editors.

PFJ through increasing of an Q-angle and as a consequence triggers of cartilage lesion [5]. It is very important , because every repairing procedure of cartilage lesions in PFJ must be predated with alignment assessment [20]. Performing cartilage reparation procedures on a limb with significant malalignment will cause treatment failure. Exact discussion on this matter exceeds assumptions of this paper.

Further diagnostic steps, especially with interpretation difficulties of performed x-rays, provides computed tomography (CT), with a possibility of using 3D reconstruction. CT apart from clarifying doubts arisen from preceded diagnostic steps also gives an opportunity to precisely measure other PFJ parameters like TTTG(Tibial Tuberosity Trochlear Groove distance), in normal knees TTTG is 13-14 mm, patellar tilt (patellar slope measured in relation to posterior femoral condyles plane) in normal knees to 20 degrees [10]. CT scan has an advantage over Magnetic Resonance Imaging (MRI) in discussed area because of a necessity of performing MRI with the knee put to the coil that imposes flexion in the knee joint. TTTG measured in the same patient in CT and MRI may differ from one another which can cause confusion when discussing border values in the context of an operative treatment [21].

MRI in comparison to CT scans is safer in a radiation aspect. Furthermore it provides good visualisation of soft tissues such as: ligaments, tendons and menisci. It enables to classify the cartilage in ICRS – International Cartilage Repair Society, proof of the presence of plica syndrome, condyles abrasion, presence of abnormal joint fluid volume and extra

articular effusion, exact localisation of free bodies especially non contrast ones (cartilaginous free body, free body in difficult localisation) and all the extra articular changes particularly in post traumatic cases [10].

Ultrasound does not have significance in diagnostic purposes in adult patients. It takes place in pediatric ones [10]. Apart from the obvious advantages such as no ionising radiation , there are few other features that makes this examination valuable [10]. In pediatric patients with no bony growth ended ultrasound provides dynamic and precise evaluation of patella position and also surrounding soft tissues [10]. This is a real time examination which can also help with doubtful cases.

Summary

Achievement of the information from an interview, physical examination, and diagnostic steps mentioned above provides a complex view of an anatomy and biomechanics of PFJ. It helps with making a difficult decision of possibility of conservative treatment or necessity of an operation. The decision, in spite of the most accurate test results stays individual to every patient and only in this matter it brings the successful treatment options both for the patient and for an orthopaedic surgeon.

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