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JOINT REPLACEMENT OF THE LOWER EXTREMITY IN PATIENTS WITH JUVENILE IDIOPATHIC ARTHRITIS

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■ **Joint replacement of the lower extremity in Juvenile Idiopathic Arthritis (JIA) is becoming more commonly performed worldwide. These young adults experience severe pain and disability from end-stage arthritis, and require joint replacement of the hip or knee to alleviate pain, and restore ambulation and function. These procedures are very challenging from the anesthesia and surgical point of view, due to small overall proportions, numerous bony and other deformities and soft tissue contractures. Joint replacement operations for JIA are best performed by experienced teams, where pre-operative and peri-operative care, and post-operative rehabilitation can be optimized in a collaborative, patient-centered environment.**

Introduction

Juvenile Idiopathic Arthritis (JIA) is a broad classification for inflammatory disease of the joints in patients who are less than 16 years of age [1, 2]. The incidence is approximately 1 : 10,000 children. JIA is an autoimmune disease of unknown etiology. Although various classifications are reported, practically speaking the disease may be oligoarticular (affecting 4 or fewer joints), polyarticular (affecting more than 4 joints but without systemic symptoms) or systemic ("Still's Disease"). Still's Disease is a systemic illness that occurs in infants and children that is characterized by fever, rash, lethargy, poor appetite and inflammation (pain and swelling) of many joints. The disease may affect the eyes (iridocyclitis), and be associated with hepatosplenomegaly, lymphadenopathy and inflammation of other organs. In general, more severe forms of JIA (and treatment) lead to stunting of overall growth.

Most of the JIA patients seen by orthopaedic surgeons for reconstructive surgery later in life have had polyarticular arthritis or "Still's Disease" many years earlier 3. They are sent to the surgeon by the rheumatologist because of recalcitrant pain, swelling, deformity, stiffness and poor ambulation. Often these patients are in their teenage years in high school or university, or early in their working career. In obtaining the history, the clinician should remember that the patient is the one with JIA, not the parent! The clinician should stand or sit directly in front of the patient, because their neck motion is often limited or painful. The history is of paramount importance. Not uncommonly, there have been interruptions in the patient's schooling, and the family situation needs some discussion regarding social support systems (especially if surgery is contemplated). The patient and their family need to be realistic with regards to the patient's physical limitations when it comes to determining later potential employment. Unrealistic expectations will lead to disappointment. Inquiry into systemic illnesses must be complete. Current and past medications should be determined, as some of these may have to be discontinued peri-operatively if they interfere with wound or bone healing, or the patient's ability to fight infection. Corticosteroid replacement therapy may be necessary during the added stress of surgery if the adrenal axis has been recently suppressed.

The physical examination must encompass a general examination and an appreciation of the movements of all relevant joints. Special attention should be paid to mobility of the neck, and whether the patient can open the mouth widely, as this will make successful anesthesia even more challenging. Gentle but thorough examination of the limbs will give the surgeon a clear picture of deformities and contractures, range of motion and stability, and the gliding potential of muscles and tendons around the joints. Assessment of the general nutritional status is important to help ensure adequate wound healing.

The surgeon should watch how the patient walks, and performs activities of daily living such as putting on their shoes and socks. Simple requests such as asking the patient to raise both upper extremities "to the sky", put their hands on their head (simulating combing of the hair), place both hands to their groin and backside (simulating toileting activities), and other simple maneuvers will give the clinician an idea of how JIA has impacted the patient's activities of daily living.

The Rheumatologist has often already performed radiographs of the joints. These should be reviewed and other radiographs ordered as appropriate, minimizing radiation exposure. For potential hip replacement patients, we obtain an anteroposterior pelvic radiograph, frog and cross table lateral. The frog lateral is particularly useful to assess the anterior femoral bow; the cross table lateral will aid in the assessment of the femoral and acetabular anteversion. For potential knee replacement patients, we obtain a standing 3-foot (91 cm) radiograph of the hips, knees and ankles on 1 long film, and lateral, notch and patellofemoral views of the knee. These are assessed for overall lower extremity alignment, deformity, degree of arthritis, bone quality, and subluxation, especially of the patello-femoral joint. Note that contractures can lead to overestimation of the size of bones due to readiographic magnification. It is useful to have a standard sized radio-opaque marker ball adjacent to the limb on the radiograph for templating purposes.

Prior to referral for hip or knee arthroplasty, the rheumatologist or another orthopaedic surgeon usually has tried different conservative treatments including physical and occupational therapy, analgesics, orthoses, anti-inflammatory and disease modifying medications. The patient may have also undergone soft tissue surgical procedures such as arthroscopy, debridement, synovectomy and tendon lengthening or releases [3, 4]. Osteotomies are often performed in younger patients with open epiphyses and deformity, with a joint that is still salvageable.

If joint replacement of the hip or knee is contemplated, the surgeon must realize that this requires a team approach to include the patient and their family, the rheumatologist, anesthesia team, nursing, social work, physical and occupational therapy, and other health care personnel. As a general guide, given end-stage arthritis of the hips and knees, we perform hip replacement prior to knee replacement, as mobile hips facilitate rehabilitation and make the knee replacement much easier to perform. All but the most simple foot and ankle surgery should follow knee replacement, as frequently the knees are aligned differently after knee replacement, thereby affecting how the ankle and foot contact the ground. This follows the general axiom of working from proximally to distally in the lower extremity, when considering surgical procedures.

Anesthesia is often very technically demanding in JIA patients. This should be dealt with by a very experienced anesthesiologist and not a novice! There is often a small, under developed mandible that is posteriorly located (retrognathia). The neck may be stiff from chronic arthritis, degenerate discs, and fusions in the cervical spine. There may be arthritis of the temporal-mandibular joint, arytenoid cartilages or other structure in the larynx. The surgeon should be patient while anesthesia is being induced! Regional anesthesia is often a preferred course, but if general anesthesia is given, this may require awake, fiber-optic intubation. All pressure points should be carefully padded during positioning of the limbs. Contractures, deformities and prominences should be accommodated and padded when operating.

Surgical technique encompasses careful pre-operative planning, meticulous surgical technique, patience and experience [5]. It is important to remember that in JIA, the bones are often small, narrow and deficient with small intramedullary canals, multi-planar deformities are common, and the soft tissues are atrophic with contractures. Because of this, the operative times are usually longer than in adults having a similar reconstructive surgery, and special instrumentation and small-sized implants are necessary.

Total hip replacement (THR) in JIA

Indications for THA include a skeletally mature patient with severe end-stage arthritis that is not salvageable by other means. Contraindications include open epiphyses, systemic or local infection, severe abductor weakness and Charcot neuropathic arthropathy.

JIA patients commonly have generalized short stature, hypoplasia of the acetabulum and femur, and excessive femoral and acetabular anteversion [6]. Standard hip components made for adults are usually too large for JIA patients. Whereas previously small components made for hip dysplasia and patients with small proportions have been cemented, more recently the trend has been to using cementless hip implants due to the fact that smaller components are now being

manufactured and theses patients may require many revisions in the future [7-18].

We have recently reported the intermediate to long-term results of THR in JIA patients 5-20 years after surgery [19]. Thirty-seven THRs in 16 women and 8 men were followed by serial clinical examinations and radiographs. The mean age of the patients was 22.6 year, with a range from 14.4 years to 35.9 years. A direct lateral approach to the hip was used in all cases. The surgical technique involved the use of off-the-shelf components including a cementless modular acetabular component and screws, combined with a cementless (26) or cemented (11) stem. The cemented stems were performed early in the series, in cases in which stable cementless fixation could not be obtained. In cementless cases, gentle flexible femoral reaming was performed prior to rasping. Two patients with 3 hips died 7 and 15 years post-operatively. Two patients with 3 well-functioning hips refused formal follow-up, but were reportedly doing well clinically. Three more patients with 4 well functioning cementless hips were lost to follow-up between 5 and 7.4 years post-operatively. Of the remaining 27 hips, 15 are intact whereas 12 (9 patients) have been revised. Of the intact hips, follow-up averaged 15.8 years (range 6.5-19.6 years). The Harris Hip Score [20] in these hips improved from an average of 48.8 (SD=20.3) to 72.5 (SD=13.5) (P=.003). Six cementless acetabular components with conventional polyethylene were revised because of wear and osteolysis after 5.5 to 14.5 years. Two of these hips had severe proximal femoral osteolysis with a well-fixed stem and had femoral bone strut grafting with wires. All 3 adult-sized Biomet cementless C2 femoral stems with minimal porous coating (used early in the series) failed. Two of 23 cementless AML Bantam stems (DePuy) loosened at 9.5 and 19.6 years. The first was undersized and the second loosened after a traumatic periprosthetic fracture. One of eight cemented AML Bantam stems (done early in the series) loosened at 3.5 years, and was associated with subsidence, cement fracture, and periprosthetic fracture with osteolysis. Pain relief and functional improvement were dramatic after total hip arthroplasty in juvenile idiopathic arthritis. We no longer cement the stems and utilize more modern bearing surfaces with highly cross-linked polyethylene in these patients.

Revision total hip replacement in JIA

We recently reported an update and extension (currently in press) of our results of revision THR in JIA [21]. This encompassed 24 revision surgeries in 15 patients with follow-up from 5-19 years. The 24 revision cases included 6 bilateral cases, 9 unilateral cases, and 3 hips that were revised twice. The indications for surgery included 22 hips with aseptic loosening and 2 hips that were re-implanted for septic loosening. A posterior approach was used in all cases, along with 2 sliding and 2 extended femoral osteotomies. Eight of the revisions were of both the acetabular and

Figure 1. Severe juvenile idiopathic arthritis (JIA) of the left hip, demonstrating joint destruction, with proximal and lateral migration of the femoral head

Figure 2. Post-operative radiograph showing a modern cementless total hip replacement

femoral components, 12 were socket revisions only and 4 femoral components alone were revised. All acetabular reconstructions involved impaction grafting of morselized cancellous bone graft, along with a cementless cup and screws (15) or a roof (3) or reconstruction (2) ring. Eight revision femoral components were cementless (1 strut graft added), and 4 cemented stems had a proximal femoral allograft (2) or strut allograft (2). Harris Hip Scores improved from 54 (range=34-85) to 77 (37-100) (p<.001). Seventy-one percent of patients reported no or slight pain post-operatively and 27% had improved functional status. At a mean 8.6 years (range 5-19 years), radiographs demonstrated that 4/20 sockets were radiographically loose (3 cementless cups, 1 reconstruction ring); two of these have been revised. Two cemented femoral stems have been revised for loosening, osteolysis and periprosthetic fracture using a long stem cementless stem and strut graft. There were 2 intra-operative femur fractures through osteolytic bone, 1 permanent sciatic nerve palsy in a patient with a reconstruction ring, 1 peri-operative and 2 late infections, and1 late dislocation. Survivorship of acetabular revisions at 9.5 years was 76%; survivorship of the stem with loosening as the end point was 80% at 9.8 years. Although the peri-operative complication rate was low, survivorship was limited by extensive bone loss, the availability of suitable implants for revision in small proportioned patients, and older conventional bearing surfaces.

Total knee replacement (TKR) in JIA

Indications and contra-indications for TKR are similar to those for THR. Severe hyperextension deformity and marked extensor weakness/ discontinuity are also contra-indications to TKA in this population.

In JIA, the knee is often very stiff due to chronic contractures and the bone is very soft due to limited weight-bearing, so careful gentle exposure with extensive soft tissue release is necessary [5, 22-24]. Quadriceps adhesions to the femur should be released to enhance gliding. The posterior cruciate ligament is always tight and this author releases it routinely, along with releasing the posterior capsule if flexion contracture is present. The metaphysis is often angulated or rotated on the diaphysis, so proper femoral bone cuts and implant alignment rely on the diaphysis, and not necessarily on the epicondylar axis or posterior femoral condyles. Bone resection should be conservative, but the aim is to get the knee into full extension. The flexion and extension gaps must be carefully balanced. The knee should not be "overstuffed" with metal and plastic so as to make subsequent motion a struggle. The patellofemoral joint is frequently very hypoplastic and mal-aligned, so that releases and guarded bone resection are necessary. Irrigation of the bone surfaces should be gentle or the soft bone will wash away. Frequently the components need to be cemented separately. Careful soft tissue handling during closure is mandatory. Post-operative range of motion exercises are begun immediately, and pain medication must be liberal, or compliance with rehabilitation will be poor. Not uncommonly, night splinting in extension will maintain a straight knee, whereas daily therapy will obtain flexion and strength.

We have recently reviewed our total knee replacements with at least 10 years of follow-up. Off-the-shelf, small total condylar or posterior stabilized knee replacements were used. The mean age of the patients was 22.4 years (range= 15-40). Five knees in 3 patients have chosen local follow-up by the family doctor, leaving 17 of 22 knees for full review. Mechanical axis was corrected from 9.4±4.2 degrees to 6.6± 5.0 degrees. The mean fixed flexion contracture improved from 29 degrees to 7 degrees (p=.001), and mean range of motion improved from 50 to 84 degrees(p=.003). One patient needed further surgery for patellofemoral re-alignment and another developed a regional pain syndrome that resolved. This small series has shown that pain relief is excellent, function and deformity improved although range of motion is only fair after TKR in JIA. In some cases, custom implants may be necessary.

Recently a large international series of TKRs performed for JIA between 1979 and 2011 by experienced surgeons at 5 hospitals were combined, including those performed by the author [25]. This comprised 349 TKRs in 219 patients, with a minimum follow-up of 2 years (mean, 12 ± 8 years; range= 2-33 years). The average age at surgery was 28.9±9.7 years (range=11-58 years). The 10-year survivorship of the TKR was 95%, however this decreased to 82% at 20 years. Revision was performed in 31 of 349 TKRs (8.9%) for primarily due to polyethylene wear and loosening, infection, stiffness, and periprosthetic fracture.

Discussion

Patients with JIA and painful, debilitating end-stage arthritis of the hip or knee have few options

Figure 3. JIA patient with bilateral hybrid total hip replacements with loosening of the right cemented femoral component and periprosthetic osteolysis

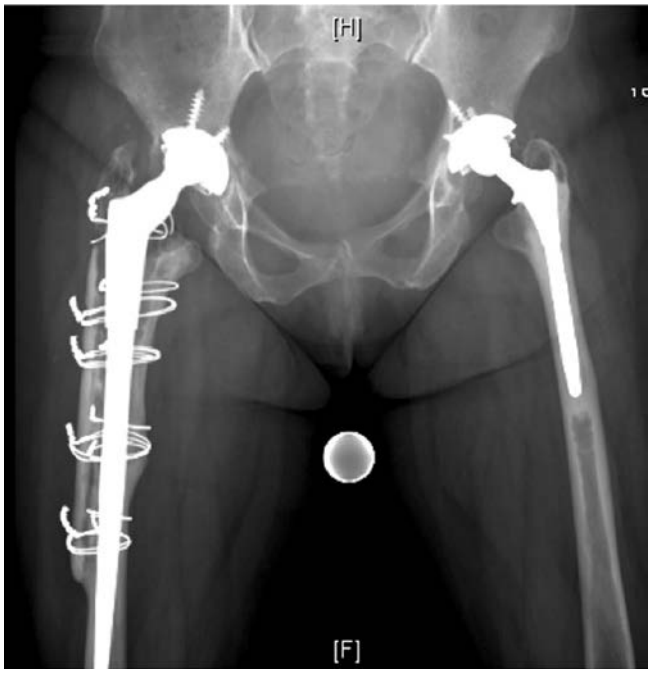

Figure 4. Ten years post revision of the right hip seen in Figure 3 with long stem cementless femoral component and strut femoral allograft

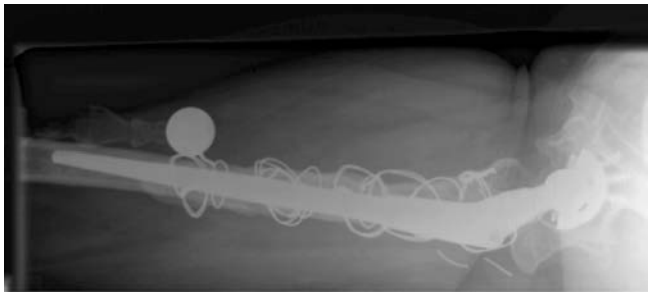

Figure 5. Cross table lateral radiograph of the right revised hip replacement

Figure 6. This same patient shown in Figures 3, 4 and 5 has had bilateral total knee replacements

other than joint replacement, if they want to remain comfortable and ambulatory. These operations are technically difficult and should be performed by experienced surgeons in a comprehensive surgical and rehabilitative setting [5, 10].

Traditionally, hip replacements were cemented in JIA, but more recently smaller cementless modular implants with different designs and sizes are being used more commonly [12-18, 26]. This provides the clinician with numerous options to re-establish more normal anatomical relationships while minimizing the potential risk of fracture or over-lengthening of the limb. Cemented stems in patients with small femoral canals left little room for the cement; modern cement technique including centralization of the stem in an adequate cement mantle was virtually impossible. This would lead to cement cracks, osteolysis, loosening and even late fracture. The surgeon should be cognizant of the excessive femoral and acetabular anteversion in JIA hips [6]. Placement of the implants must provide a stable range of motion without subluxation or dislocation.

Total knee replacement provides even more challenges in JIA. Due to chronic inflammation and subsequent abnormalities in growth, the femoral condyles are often large, externally rotated and posteriorly located in relation to the shaft [22, 24]. This makes standard sized knee replacements difficult to position accurately and fit onto the bony surfaces. Post-inflammatory fibrosis leads to contractures and adhesions that do not facilitate the gliding of soft tissues over the prosthesis. Balancing the flexion and extension gaps is also challenging, and will lead to difficulties in range of motion or instability if not accurately performed. The bone is soft and care must be taken to prevent intra-operative fracture or crushing of the metaphyseal bone. Because of these challenges, meticulous, gentle technique should be followed, and components cemented. Clinical outcomes, previously very guarded, seem to be improving with newer more anatomical implants [25, 27-31].

Figure 7. JIA patient with bilateral total hip replacements has severe arthritis of both knees

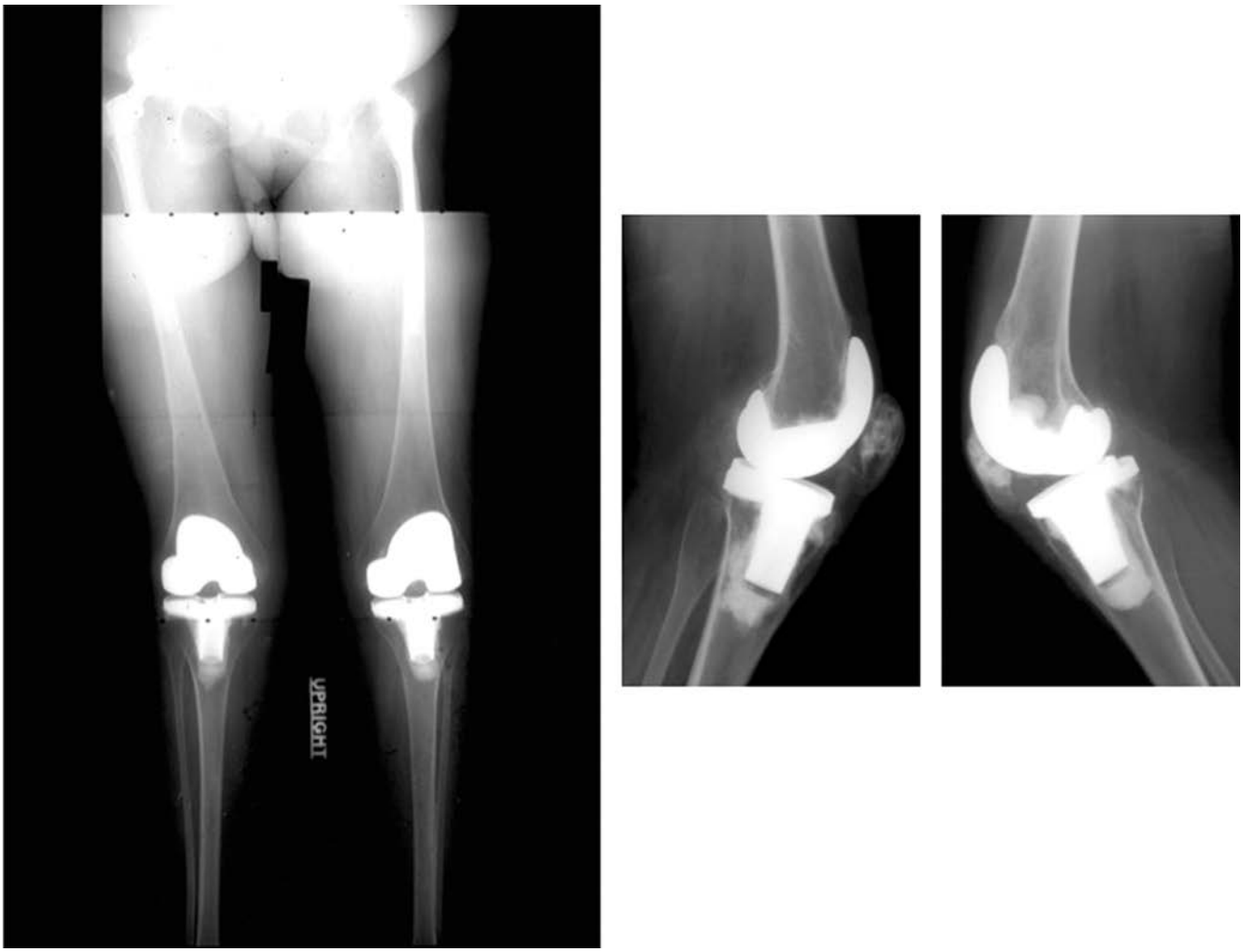

Figure 8. Post-operative radiographs after bilateral total knee replacement

Hip and knee replacements have the potential to change the lives of young adults afflicted with JIA. Continued improvements in prosthetic design, bearing materials and surgical technique offer hope that implant longevity will be further extended and more normal function restored.

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ЭНДОПРОТЕЗИРОВАНИЕ СУСТАВОВ НИЖНИХ КОНЕЧНОСТЕЙ У ПАЦИЕНТОВ С ЮВЕНИЛЬНЫМ ХРОНИЧЕСКИМ АРТРИТОМ

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■ **Эндопротезирование суставов нижних конечностей у пациентов с ювенильным хроническим артритом становится все более распространенной операцией во всем мире. Эти пациенты уже в молодом возрасте страдают от тяжелого болевого синдрома и двигательных ограничений вследствие необратимых изменений в суставе и нуждаются в замещении тазобедренных и коленных суставов эндопротезом для купирования болевого синдрома и восстановления двигательных функций. Эндопротезирование в таких условиях представляет собой сложную задачу как с анестезиологической, так и с хирургической точки зрения вследствие общих малых размеров костей, их деформаций и контрактуры мягких тканей. Эндопротезирование суставов у пациентов с ЮХА нужно проводить при наличии опытной многопрофильной команды специалистов, обеспечивающей оптимальное пре-, интра- и послеоперационное ведение, преемственность и ориентированность на наилучший для пациента результат.**

Уважаемые коллеги!

Для меня большая честь опубликовать свою статью «Эндопротезирование суставов нижних конечностей у пациентов с ювенильным хроническим артритом» в новом и многообещающем российском журнале «Детская травматология, ортопедия и восстановительная хирургия». Эта статья обобщает мой более чем 25-летний опыт лечения ювенильного хронического артрита в Стэнфордском университете. Эндопротезирование суставов в целом интенсивно развивалось в течение последних десятилетий, соответственно расширялись возможности лечения молодых пациентов с артритом, обеспечивая стойкое облегчение боли и улучшения функций.

Написание этой статьи связано с лекцией, которую я прочитал в Детском ортопедическом институте имени Турнера в Санкт-Петербурге в июне 2013 г. По окончании лекции состоялась плодотворная дискуссия с коллегами. Мой друг, доктор Владимир Кенис, помогал мне во время визита и перевел статью на русский язык. Я надеюсь, что эта статья откроет долгосрочное и полезное сотрудничество между нашими клиниками. Я поздравляю вас с выходом нового журнала, который должен помочь в этом.

С наилучшими пожеланиями,
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