

Severe Disease Activity and Complications of Immunosuppressive Therapy: A Challenge for Acute Hospital-based Rehabilitation in Rheumatology

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ABSTRACT. Acute rehabilitation refers to the multidisciplinary rehabilitative treatment of patients in continuing need of integrated acute and rehabilitative longterm care. As a result of the advances in acute rheumatology and improved emergency services, an increasing number of patients survive episodes of severe disease and complications of immunosuppressive therapy. These patients require not only treatment of their acute medical problems but also specialized multidisciplinary acute rehabilitation starting as early as possible during their hospital stay. We describe 4 typical cases from the major fields of rheumatology. (1) Acute rehabilitation of a 63-year-old woman with rheumatoid arthritis after replacement of both preexisting knee endoprostheses in one session and removal of the left hip endoprosthesis due to infection and sepsis. (2) Rehabilitation of a 29-year-old man with a 7-year history of ankylosing spondylitis who lived in an adjustable easy chair for 2 years due to severe pain prior to admission. (3) A 61-year-old woman with active refractory Wegener's granulomatosis who developed respiratory insufficiency due to aspergillus and pseudomonas pneumonia. (4) The acute rehabilitation of a 21-year-old woman with systemic lupus erythematosus and a history of 14 laparotomies due to severe acute pancreatitis and multiple gut perforations. Acute rehabilitation was complicated by a large defect of the abdominal wall and significant critical illness polyneuropathy. Our report points out differences between acute, postacute, and longterm rehabilitation, describes the mobilization of patients in acute rheumatology units, and defines specific problems encountered in acute hospital-based rehabilitation of rheumatological patients. (First Release June 15 2009; J Rheumatol 2009;36:1618–25; doi:10.3899/jrheum.081136)

Key Indexing Terms:

REHABILITATION

QUALITY OF CARE

PHYSICAL THERAPY

RHEUMATOID ARTHRITIS

ANKYLOSING SPONDYLITIS

WEGENER'S GRANULOMATOSIS

SYSTEMIC LUPUS ERYTHEMATODES

As a result of ongoing advances in acute rheumatology and improved emergency services, an increasing number of patients survive episodes of severe disease and complications of immunosuppressive therapy. Such patients require not only treatment of their acute medical problems but also specialized multidisciplinary acute rehabilitation starting

very early during the hospital stay. Acute rehabilitation refers to intensive efforts involving therapy and reeducation to help a patient regain mobility, strength, and flexibility following a severe injury^{1,2}. Any acutely hospitalized individual who has a new disability or exacerbation of an existing one is a candidate for acute rehabilitation. This can vary from something as straightforward as weakness-related inability to walk or perform activities of daily living (ADL), to new swallowing difficulties, to higher-level thinking deficits. Typically, once patients are admitted to the rehabilitation process, rehabilitation professionals assess their functional status to evaluate their potential for rehabilitation and to develop a care plan³.

Today, 3 phases or settings of medical rehabilitation are distinguished: acute rehabilitation or very early rehabilitation; post-acute rehabilitation; and longterm rehabilitation. Acute rehabilitation can be performed in specialized units for acute rehabilitation (ARU) led by a specialist or a mobile team working in an acute-care hospital.

Depending on the kind of acute rehabilitation needed by the patient, there are differences in intensity and amount of

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therapies. Acute rehabilitation performed in a discrete, licensed unit (ARU) is defined as rehabilitation with at least 3 hours of daily therapy and 2 different types of therapy (high intensity), whereas mobile therapy is performed with 0.5–1.5 hours a day, 5–7 days a week (moderate intensity) and with at least 1 therapy type.

The ARU of the Department of Physical and Rehabilitation Medicine (PRM) of Charité–Universitätsmedizin Berlin consists of a 7-bed ward. The PRM department provides all therapeutic facilities needed to provide medical care and state of the art rehabilitation services. The therapeutic team is headed by specialists in physical and rehabilitation medicine and consists of physicians, physiotherapists, occupational therapists, and psychologists.

Physical therapy is focused on mobility skills (e.g., transfers, wheelchair use, walking), leg flexibility and strengthening, trunk control and balance, endurance training, and using adaptive equipment to facilitate mobility. The occupational therapist's work is focused on activities of daily living skills (feeding, dressing, bathing, grooming) and using adaptive equipment to facilitate ADL.

A mobile rehabilitation team led by a physician specializing in physical and rehabilitation medicine is available to start acute rehabilitation while patients are still undergoing treatment on the different wards for their medical problems. In all settings of acute rehabilitation, nursing care is an integral part of any rehabilitation program and good teamwork is essential for optimal patient care. Therefore, most members of the nursing team have been trained in kinesthetics and some have had special training for rehabilitation care.

In a weekly team meeting, the staff providing ARU care and the members of the mobile team discuss the most suitable rehabilitation strategy for an individual patient from different points of view. This results in a comprehensive rehabilitation regimen tailored to the individual patient and recorded in a plan that defines the next goals to be achieved by the patient. Further, rehabilitation measures are integrated into the individual patient's acute medical treatment plan. The aim is to improve the patient's reduced functional capabilities, which comprise body functions and structures, and to restore health to the extent that the patient can undergo treatment in a postacute or longterm rehabilitation facility after the acute hospital stay.

Rehabilitative measures offered by a multidisciplinary therapeutic team similar to the acute rehabilitation team described above are implemented in the German Diagnosis-Related Group system. There is general agreement by now that an integrated therapeutic strategy is necessary for successful treatment of patients with rheumatic disorders^{4,5}.

In rheumatology, acute rehabilitation mostly refers to rehabilitative interventions at an early stage of disease⁶. In general, acute rehabilitation describes the multidisciplinary treatment of patients in continuing need of integrated acute and rehabilitative care. It means the earliest possible initia-

tion of rehabilitation combined with the necessary acute medical care of patients provided by various specialties⁷. The risk of a significant loss of function is increased in patients with long stays in intensive care units (ICU), critically ill patients, and patients with preexisting chronic conditions⁸. Therefore acute rehabilitation is defined as a procedure in the Diagnosis-Related Group system and comprises specialized neurological, geriatric, and multidisciplinary care⁹.

The aim of this article is to exemplify the challenges facing acute rehabilitation in rheumatology by describing 4 cases from the major fields of rheumatology, namely rheumatoid arthritis (RA), ankylosing spondylitis (AS), vasculitis, and systemic lupus erythematosus. The focus is on deficits in body function of patients who have survived episodes of severe disease or complications of immunosuppressive therapy and the enormous therapeutic input required in the form of individual therapeutic sessions from various disciplines involved in rehabilitation. All 4 patients underwent rehabilitation in the ARU of our department. During their medical treatment before referral to the ARU, rehabilitation was performed by mobile teams.

Acute Rehabilitation Cases from the Major Fields of Rheumatology

Case 1: Rheumatoid arthritis. With the resources of modern acute rheumatology it is possible to control high disease activity in RA. It is prolonged recovery from endoprosthetic replacement of the large joints or severe complications of immunosuppressive therapy or both that brings a patient in need of integrated acute and rehabilitative care. In longstanding RA, normal mobilization and rehabilitation are limited by the preexisting destruction and glucocorticoid-induced myopathy.

A 63-year-old woman with a 20-year history of RA (Figures 1A, 1B), endoprosthetic replacement of both knees in 1994-95 and the left hip in 2004, and spondylodesis of C0/C4 in 2001 and T12-L5 in 2000 (Figures 1C, 1D) was under treatment with methotrexate and leflunomide in combination with glucocorticoids (15 mg/day for more than 3 years). She was admitted directly to the ICU with respiratory insufficiency and renal failure due to sepsis caused by *Staphylococcus aureus* infection of the endoprostheses of both knees and the left hip. She underwent one-stage knee endoprosthesis exchange and creation of a Girdlestone situation in the left hip joint (Figures 1E-1G). After orthopedic intervention she was treated in the ICU for 12 days, followed by another 10 days on a sterile ward, before she was transferred for multidisciplinary rehabilitation to the ARU. The clinical examination revealed generalized muscle atrophy, reduced strength of the arms and legs (3/5 according to Janda of the arms and 2/5 of the legs). She suffered from glucocorticoid-induced myopathy and critical illness polyneuropathy. When she was transferred to our unit, she

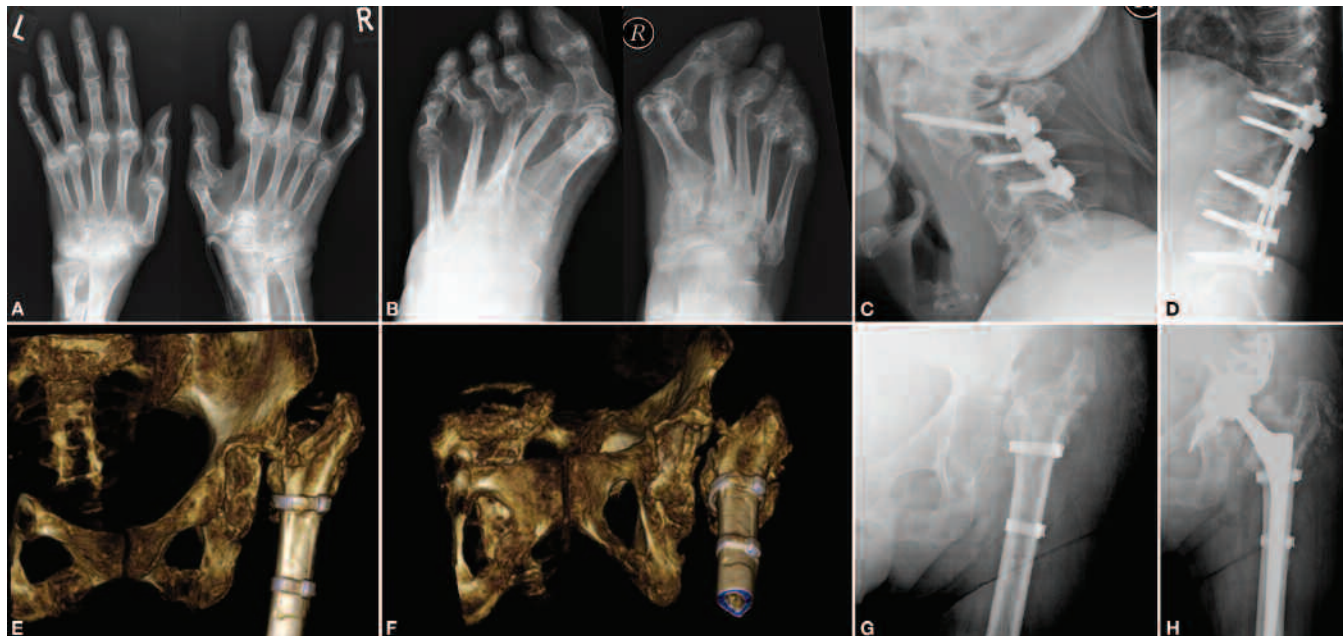


Figure 1. A 63-year-old woman with rheumatoid arthritis. Radiographs of hands (A) and feet (B) show severe joint destruction and deformity. C. Status post C0-C4 stabilization. D. Status post T12-L5 spondylodesis. E, F. 3-D reconstructions of a computed tomography scan of the left hip showing Girdlestone situation and titanium cerclage of the fractured femur. G. Cranialization and demineralization of the Girdlestone femur. H. Radiograph after left hip endoprosthesis reconstruction with a Burch-Schneider ring (socket).

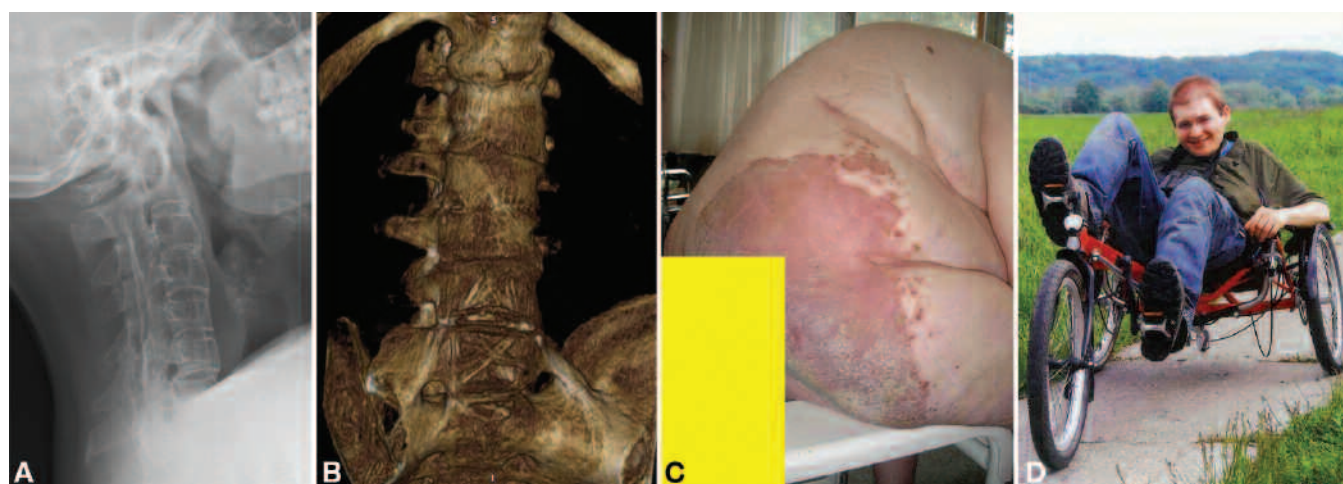


Figure 2. A 29-year-old man with ankylosing spondylitis. A. Lateral radiographic view of the cervical spine showing bamboo spine. B. 3-D reconstruction of a computed tomography scan of the lumbar spine and sacroiliac joints showing severe ankylosis. C. The patient on admission showing extreme obesity and psoriasis. D. The patient today.

could not sit without support, stand, or walk. Transfer was not possible without assistance. She could not eat without assistance because she was unable to move her hands toward her mouth. The post-event Barthel index^{10,11} was only 20 points. Before the septic event, she had been completely independent and did not need any assistance in daily living. Due to the marked loss of function, with reduced ADL, she experienced reactive depression. The aim of acute rehabilitation in this patient was to achieve stabilization to an extent

that she could undergo surgical restoration of the left hip endoprosthesis. Her treatment consisted of daily fractionated individual therapy sessions, including physical exercises on a neurophysiological basis for stabilization of the trunk several times a day, transfer training assisted by 2 physiotherapists, respiratory therapy for pneumonia prophylaxis and to restore normal breathing, and occupational therapy for training in ADL. Altogether, she received 3–4 hours of individual training per day 7 days a week. The physicians

caring for her included a specialist in physical and rehabilitation medicine, a rheumatologist, a pain therapist, and a psychiatrist. After 8 weeks of acute rehabilitation, she was able to undergo surgery and the planned hip endoprosthesis restoration was performed, including stabilization of the socket with a Burch-Schneider ring (Figure 1H). Postoperatively, a second phase of acute rehabilitation was indicated; the aim was to restore a partial loading capacity of 15 kg of the left leg. She received individual sessions with the same frequency as preoperatively to train in standing and walking, first with a hydraulic walking device, then with a walking frame. At the end of acute rehabilitation, she was able to transfer without assistance and had a walking distance of 20 meters with the hydraulic device and 10 meters with a walking frame. The Barthel index increased to 35 points at discharge. She was discharged to home rehabilitation after acute and post-acute rehabilitation was concluded.

Case 2: Ankylosing spondylitis. A 29-year-old man (Figure 2A-2D) with a 7-year history of AS and endoprostheses of the hips lived in an adjustable easy chair for 2 years before admission to the hospital due to severe back pain. He left the chair only when his parents transferred him to the bathroom. He was treated with methotrexate, sulfasalazine, glucocorticoids, and nonsteroidal antirheumatic drugs. We selected this case because of the high disease activity, which led to complete immobility.

On admission, he had fixed hyperkyphosis of the thoracic spine, extreme obesity (weight 115 kg, height 177 cm), and extensive psoriatic skin lesions of the trunk and in the genital area (Figure 2C). Radiography demonstrated the typical changes of AS in the cervical spine (Figure 2A). There was complete ankylosis of the sacroiliac joints on both sides (Figure 2B).

Treatment in the acute hospital setting was necessary because of the high level of impairment and also because strict monitoring of side effects of a new drug regimen was needed. Further, daily intensive skin care was performed in the department of dermatology.

He was admitted to the ARU because of functional loss and inability to perform ADL due to high disease activity and severe pain. In spite of intensive physical and occupational treatment, psychological therapy, and pain management, no substantial improvement was achieved due to the high activity of AS. The spinal form of psoriatic arthritis was discussed as a differential diagnosis. After initiation of treatment with the tumor necrosis factor- α inhibitor infliximab, an impressive reduction of disease activity and pain was observed. The full acute rehabilitation program with 6 hours of individual therapy could be started. The impressive gain in function achieved in this patient was only possible with the early onset of an integrated acute rehabilitation program. The aim of the program for this patient was to restore function to an extent that he could undergo further post-acute rheumatological rehabilitation on an outpatient

basis (Figure 2D). This was achieved after 57 days of acute rehabilitation and a total of 97 days as an inpatient. He was able to do the transfers by himself and walked short distances with a hydraulic walking device. The Barthel index increased from 30 points on admission to 65 points at discharge from the ARU.

Case 3: Vasculitis. To illustrate the severe complications that may be associated with modern immunosuppressive therapy, we describe a 61-year-old woman with a 5-month history of high activity Wegener's granulomatosis (Figures 3A-3C). Despite cyclophosphamide and glucocorticoid pulse therapy, followed by oral cyclophosphamide accompanied by high-dose glucocorticoids (up to 60 mg), the vasculitis activity remained high, with bronchial stenosis and mononeuritis multiplex resulting in paresis of the facial, glossopharyngeal, and recurrent nerves. Only B cell depletion with rituximab controlled the disease. One month after discharge from the hospital, the woman was hospitalized with severe respiratory insufficiency due to pneumonia (*Aspergillus* and *Pseudomonas* infection) and recurrent highly active vasculitis with new bronchial stenosis (Figure 3B). The 5-month course of high-dose glucocorticoids led to severe osteoporosis with fractures of the T8-T10 vertebrae (Figure 3D). Vertebroplasty was performed for pain management, but was complicated by embolism of a pulmonary artery (Figures 3E-3G), and the patient's respiratory situation worsened. The complicated pulmonary situation with bronchial stenosis and mucus retention and reduced mobility due to severe back pain was the indication for transferring her to the ARU. Her treatment comprised intensive physical respiratory therapy including reflectory respiratory treatment several times per day to reduce mucus retention in order to prevent a new systemic infection with *Pseudomonas*, which was multiresistant by then.

The aim was to improve her condition to the point that she could be referred for further post-acute rehabilitation in a pulmonary facility, and this was achieved after 19 days of acute rehabilitation and a total hospital stay of 149 days. The Barthel index increased from 50 points on admission to 85 points at discharge from the ARU.

Case 4: Systemic lupus erythematosus. A 21-year-old woman had systemic lupus erythematosus with multiple gut perforations and severe acute pancreatitis. Acute treatment consisted of modified extended hemicolectomy on the left with blind closure of the descending colon, terminal stoma of the ascending colon, and lavage. The course was complicated by sepsis and multiple organ failure. Repeated laparotomies were needed because of abscess formation (Figure 4A).

Mobile rehabilitation was started right after ICU treatment with 48 days of artificial respiration and a total of 14 laparotomies. A large defect of the abdominal wall covered only by an artificial mesh (Figures 4B-4E) and significant critical illness polyneuropathy complicated the course of acute rehabilitation.

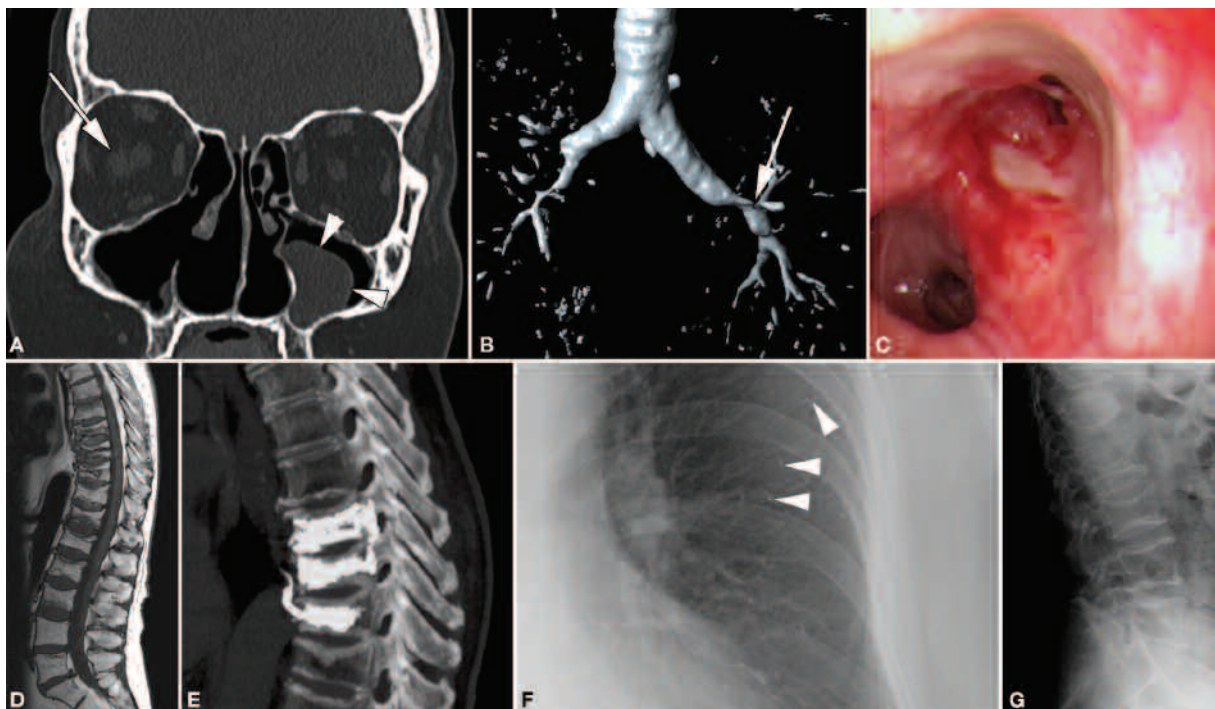


Figure 3. A 61-year-old woman with a 5-month history of Wegener's granulomatosis. A. Granulomas in the paranasal sinuses (arrowheads) and in the orbit (arrow). B. 3-D reconstruction of a computed tomography scan of the lungs showing stenosis of the right main bronchus (arrow). C. Fiberoptic bronchoscopic view showing steroid-induced vertebral fractures of T8-T10. D. Magnetic resonance imaging scan showing steroid-induced vertebral fractures of T8-T10. E. Status post-vertebroplasty; displacement of bone cement into spinal canal and paravertebral veins shown by computed tomography (sagittal view). F. Magnified chest radiograph shows pulmonary artery embolism caused by bone cement (arrowheads). G. Lateral radiograph of the lumbar spine showing additional impression fractures.

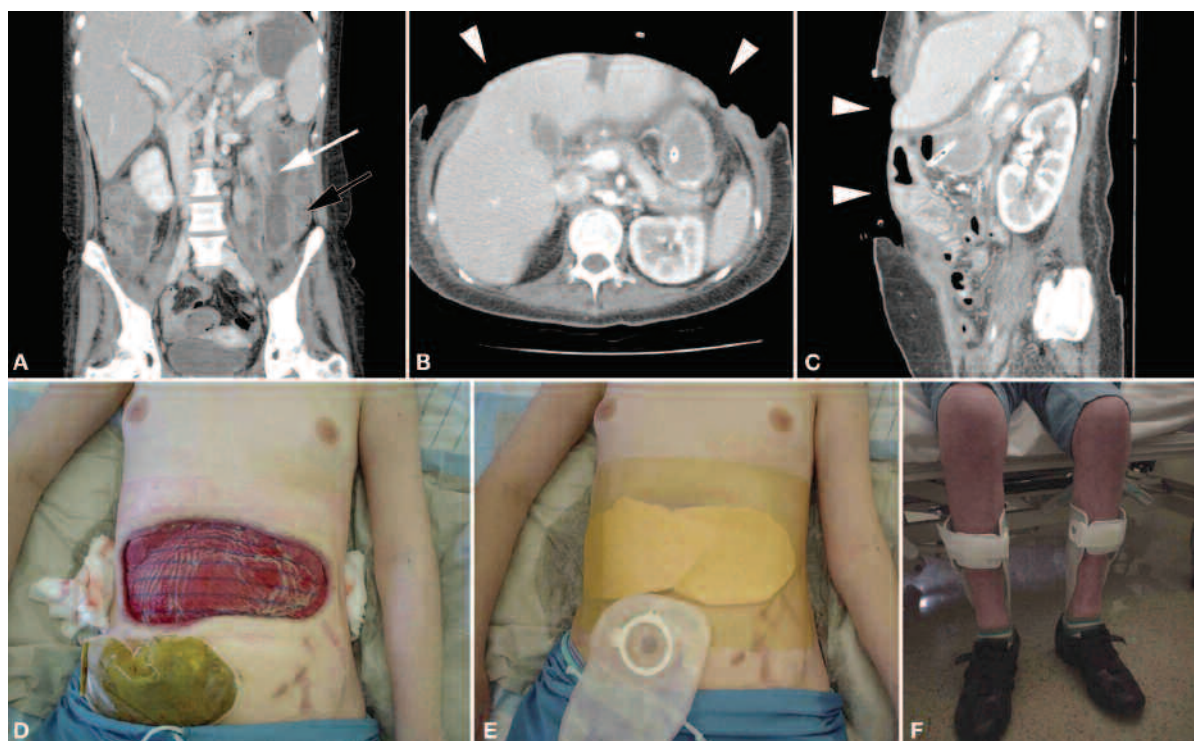


Figure 4. A 21-year-old woman with systemic lupus erythematosus. A. Abscess (white arrow) adjacent to the descending colon (black arrow). Transverse (B) and sagittal (C) images of abdominal wall defect (arrowheads) after repeat laparotomy. D. Abdominal wall defect covered with an artificial mesh and colostomy with only a small tissue bridge between. E. Abdominal wall defect with bandage. F. Livedo reticularis, peroneal splints (critical illness polyneuropathy).

When transferred to the ARU, she was able to walk a few steps with a hydraulic walking device. Unassisted transfer was not possible. Her Barthel index was 45 points. Acute rehabilitation comprised at least 4 treatment units per day with physical exercises and walking training with the hydraulic walking device, later with a walking frame and then forearm crutches. She received peroneal splints due to peroneal nerve palsy secondary to critical illness polyneuropathy (Figure 4F). The trunk was additionally stabilized with an abdominal bandage. Continual rheumatological monitoring was necessary. In addition, she received occupational therapy, pain therapy, and psychiatric and psychological treatment. A physician and a specialist nurse changed the bandage once a day. The patient started to learn stoma care. The primary aim of acute rehabilitation, early mobilization and mental stabilization, was achieved after 77 days. She had a Barthel index of 85 points when she was discharged from our ward. Because of *de novo* lupus activity, she was transferred back to the acute rheumatology ward to optimize antirheumatic drug therapy. One week later she was discharged home.

Time Requirements for Acute Rehabilitation

According to the guidelines of acute rehabilitation in Germany the necessity of acute medical care and the extent of functional loss due to disability or impairment are the admission and discharge criteria for acute rehabilitation. A widely used instrument for measuring functional impairment is the Barthel index.

In our multidisciplinary ARU, patients are usually treated until they have reached a functional level that they can undergo post-acute and longterm rehabilitation, i.e., they have a Barthel index > 70 points. Patients undergo acute rehabilitation typically after prolonged stays in the ICU with gastroenterological, cardiological, nephrological, and oncological underlying diseases, and also with sepsis and multiple organ failure, management after orthopedic operations, in particular endoprosthesis replacement surgery, and patients with functional deficits after stroke. The average time spent in the ARU was 37.9 days (standard deviation 10.1 days, range 9–106 days) between July 2006 and June 2007 (n = 143).

The hospital stays of 3 of the 4 rheumatological patients described here were markedly longer than the average (Table 1), because the patients had conditions with high disease activity or complications of immunosuppression, and the longer hospital stays reflect the complexity of acute rehabilitation. In these 4 cases, acute rehabilitation accounted for 13% to 67% of the time these patients spent in hospital. The functional features of all 4 patients on admission and at discharge from the ARU are summarized in Table 2.

DISCUSSION

In rheumatology, severe functional impairment resulting from acute exacerbation often affects patients who already have functional deficits from their underlying disease. Patients with sepsis who spend prolonged time in the ICU are likely to have loss of muscle strength due to critical illness polyneuropathy. In our department, 50% of all patients referred for acute rehabilitation after ICU treatment over a period of 2.5 years had critical illness polyneuropathy¹². Patients with a long history of a rheumatological disease often also have glucocorticoid-induced myopathy^{13,14}.

The most important difference between transdisciplinary acute rehabilitation and complex multimodal treatment of patients with acute rheumatological diseases consists in the use of staff-related therapeutic resources, the intensity of treatment, and the range of therapies offered. Complex multimodal rheumatological treatment comprises at least 3 of the following areas: physical therapy, occupational therapy, pain therapy, cognitive behavioral therapy, and speech therapy. These are combined with a total of 11 hours per week under the supervision of a rheumatologist.

Transdisciplinary acute rehabilitation requires that at least 4 of the following areas are offered: physical therapy/exercising, occupational therapy, neuropsychology, psychotherapy, logopedics/facio-oral therapy/speech therapy, dysphagia therapy, and art therapy, and that at least 3 of these are combined and performed in 15 to 40 treatment units, each consisting of at least 30 minutes, under the supervision of a physician specializing in physical and rehabilitation medicine⁹. Another feature is rehabilitative care aimed at promoting the patient's competence in activities of daily living.

Table 1. Time requirements for treatment of the 4 selected cases.

	Case 1, RA	Case 2, AS	Case 3, WG	Case 4, SLE
Total time in hospital, days	159	97	149	139
Acute medical care*, days	53 (10 ICU)	40	130	62 (56 ICU)
Acute rehabilitation, days	106	57	19	77
Percentage of time requirement for acute rehabilitation	67	58	13	55

* Intensive care unit (ICU)/rheumatology/orthopedic ward. RA: rheumatoid arthritis; AS: ankylosing spondylitis; WG: Wegener's granulomatosis; SLE: systemic lupus erythematosus.

Table 2. Functional indications on admission and at discharge.

	Case 1, RA	Case 2, AS	Case 3, WG	Case 4, SLE
Referred from	Infectiology	Rheumatology	Infectiology	ICU
Barthel index				
On admission	20	30	70	45
At discharge	35	65	90	85
Individual therapy sessions per day	4-5	6	3-4	4
Mobility on admission/ADL	Cannot walk, unassisted sitting not possible, no transfer, assistance with all ADL	Cannot walk, eating without assistance, help with grooming	Can walk a few meters, eating without assistance, help with grooming	A few steps with a hydraulic walking device, eating without assistance, help with grooming
Mobility at discharge/referral	Walking not possible, can sit and eat without assistance	Can get up without assistance, can walk a few meters with a hydraulic walking device	Can do transfers, walks 50 meters with walking frame, short distances with wheelchair	Walks 100 meters with forearm crutches
Specific treatment	Physiotherapy on neuro-physiologic basis, respiratory training, transfer training with 2 therapists, occupational therapy	Physiotherapy, physiotherapy in water with 2 therapists, occupational therapy	Respiratory therapy, reflectory respiratory therapy, inhalations, individual physiotherapy, gait training	Physiotherapy, gait training using the hydraulic walking device and walking frame, stoma care
Other treatment	Pain therapy, psychiatry	Psychosomatics, pain therapy, dermatology, rheumatology	Infectiology/pulmonology, rheumatology	Psychiatry, psychology, rheumatology
Discharge	To home, scheduled for further orthopedic surgery, and home health rehabilitation	For further rehabilitation (post-acute rehabilitation)	To home, further rehabilitation planned (post-acute rehabilitation)	To home, further rehabilitation planned (post-acute rehabilitation)

Abbreviations as for Table 1.

Chronic inflammatory rheumatic disease and multiple structural or functional damage was identified as a constellation that requires acute rehabilitation by a German expert group on acute rehabilitation⁷.

Affected patients are often young, with protracted courses of rheumatic disease and long hospital stays despite intensive management and repeated phases of acute rehabilitation. These patients require numerous diagnostic tests including close monitoring of disease activity and complex multidisciplinary management. These 4 cases illustrate the need for acute rehabilitation that goes beyond complex multimodal rheumatologic therapy. Systematic reviews of the literature provide strong evidence that multidisciplinary rehabilitation is efficient in patients with RA¹⁵. Early identification of rehabilitation needs and early initiation of rehabilitation can lower healthcare costs by reducing dependence and nursing care, shortening hospital stays, and preventing disability⁸.

In summary, acute rehabilitation is indicated in patients who develop severe functional deficits due to severe active rheumatic disease or due to multiple organ failure as a complication of immunosuppressive medication, leading to longterm ICU treatment and the development of critical illness polyneuropathy.

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