

Patients with RA-ILD were more likely to have used steroids than were RA controls (59% vs 14%;  $p=0.01$ ). Patients with RA-ILD were also more likely to have UIP ( $p=0.02$ ). The excess of this ILD subtype may account for some of the increased mortality.

Conversely, the BRILL Network has recently reported that certain immunosuppressants and biologic therapies appear to carry a survival benefit.

Since all these data are retrospective, however, 2 prospective therapeutic trials are planned. The first will recruit RA patients with progressive ILD and compare the use of mycophenolate mofetil and azathioprine, allowing for the use of oral steroids. The second will recruit patients with active RA and ILD and compare the potential survival advantage of using rituximab over anti-tumor necrosis factor- $\alpha$  therapies.

Concluding, Dr. Kelly observed that patients with RA and concomitant ILD have increased mortality compared with controls and that more than half of these patients have been treated with oral steroids. This large, retrospective, multicenter study suggests that the long-term use of oral steroids should be reconsidered when possible in RA patients who also have ILD.

## Long-Term Adalimumab Is Safe and Achieves Disease Control in Children With Advanced and Refractory JIA

Written by Jenny Powers

Long-term disease control was observed with adalimumab administered in routine clinical practice to patients with moderately to severely active juvenile idiopathic arthritis (JIA), according to Gerd Horneff, MD, Asklepios Klinik Sankt Augustin, Sankt Augustin, Germany.

Interim findings from an analysis done at 4 years were reported from an ongoing international registry of patients enrolled upon diagnosis of moderately to severely active JIA [NCT00783510]. The multicenter, noninterventional observational study aimed to evaluate long-term safety and effectiveness of adalimumab, which was recently approved for treating patients with severe and refractory JIA from the age of 2 years after failure or intolerance to methotrexate (MTX) treatment. Efficacy and safety were compared with MTX treatment administered in routine clinical practice. Patients aged 2 to 17 years were enrolled in the United States, European Union, and Australia. The planned follow-up was 10 years after enrollment in either treatment arm; cross-over to the adalimumab group was allowed.

Patients in the adalimumab cohort had a more advanced disease course compared with those of the MTX cohort. Baseline values for mean disease duration were 1.3 years in the MTX arm and 3.8 years in the adalimumab arm. Patients in the adalimumab arm had a mean age of 12.2 years, a mean weight of 47.8 kg, and a mean height of 150.3 cm compared with patients receiving MTX, who had a mean age of 9.6 years, a mean weight of 37.9 kg, and a mean height of 137.2 cm. Mean baseline active joint count in both groups was 5.8, and disability index of Childhood Health Assessment Questionnaire scores was 0.6 in each arm.

At data cutoff, 306 patients received MTX monotherapy, and 459 received adalimumab with or without MTX. Clinical outcomes based on the 71-joint Juvenile Arthritis Disease Activity Score showed that mean scores improved from 13.1 at baseline to 11.2, 6.4, and 5.1 with MTX and from 12.1 at baseline to 8.5, 5.7, and 5.4 with adalimumab at Months 1, 3, and 6, respectively. In the MTX arm, 131 (42.8%) patients discontinued registry participation, compared with 81 (17.6%) in the adalimumab arm; 34 (25.9%) who discontinued MTX were switched to adalimumab.

Discontinuation due to treatment-related adverse events (AEs) occurred in 23 (7.5%) and 25 (5.4%) patients in the MTX and adalimumab cohorts, respectively. AEs determined by the investigators as possibly drug related were reported in 63 (20.6%) patients. Infectious AEs were reported for 64 (20.9%) and 80 (17.4%) of patients in the MTX and adalimumab arms, respectively. Of these, serious infectious AEs occurred in 7 (2.3%) MTX patients, compared with 12 (2.6%) adalimumab patients (Table 1).

Table 1. Overview of Observational Adverse Events, n (%)

Adverse Event	Methotrexate (n=306)	Adalimumab (n=459)
Any	125 (40.8)	142 (30.9)
At least "possibly drug related" per the investigator	63 (20.6)	71 (15.5)
Severe	12 (3.9)	22 (4.8)
Serious	16 (5.2)	30 (6.5)
Leading to discontinuation of study drug	23 (7.5)	25 (5.4)
Infectious	64 (20.9)	80 (17.4)
Serious infectious	7 (2.3)	12 (2.6)
Injection site related	3 (1.0)	20 (4.4)

In the adalimumab arm, 12 (2.6%) patients had a serious infectious AE: acute tonsillitis, cellulitis,



gastroenteritis, mononucleosis, viral meningitis, pneumonia, scarlet fever, subcutaneous abscess, tonsillitis, urinary tract infection, and varicella occurred in 1 patient each. No deaths, malignancies, tuberculosis, or opportunistic infections were reported with either treatment.

## Aneurysm Size Does Not Predict Dissection or Rupture in GCA

Written by Toni Rizzo

The aorta and its branches are involved in about 30% of patients with giant cell arteritis (GCA) [Nuenninghoff DM et al. *Arthritis Rheum* 2003]. Patients with GCA have a 17-fold risk for developing a thoracic aortic aneurysm and a 2.4-fold risk for developing an abdominal aneurysm [Evans JM et al. *Ann Intern Med* 1995]. Aortic aneurysm in GCA is poorly understood, with few data on risk and predictors of dissection and rupture. The current guidelines for monitoring and management of aortic aneurysm do not address this unique population.

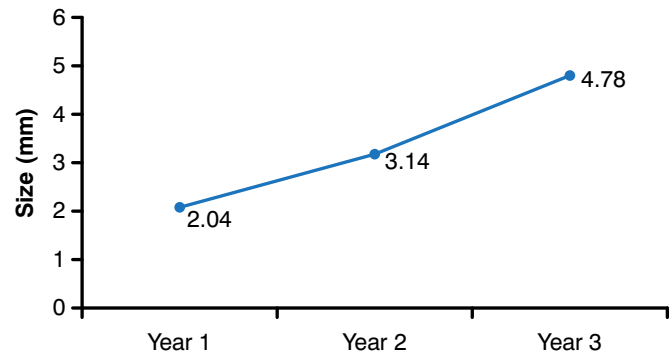
This retrospective study, presented by Ashima Makol, MD, Mayo Clinic, Rochester, Minnesota, USA, had 3 objectives: (1) to systematically study aortic dissection in patients with GCA with aortic aneurysms, (2) to describe the clinical features and outcomes of aortic dissection in patients with GCA, and (3) to determine the average growth rate of aortic aneurysms and identify aneurysm size associated with aortic dissection.

The study involved patients with GCA diagnosed at the Mayo Clinic between 2000 and 2012. Data were collected from the records of patients diagnosed with GCA and aortic aneurysms with aneurysm size measured on aortic imaging. Abstracted data included clinical characteristics at diagnosis of aneurysm, aortic aneurysm dimensions at diagnosis and on follow-up imaging, and clinical presentation at the time of dissection or rupture.

A total of 195 patients with GCA and aortic aneurysms were identified, 161 (82%) in the ascending thoracic aorta, 21 (11%) in the descending thoracic aorta, and 13 (7%) in the abdominal aorta. The mean patient age was 74 years, and 62% of patients were women. The overall mean aortic size at diagnosis was 49.3 mm. The average aneurysm overall growth rate was 1.59 mm/year for the first 3 years after diagnosis. Growth rates at 1, 2, and 3 years from baseline were 2.04, 3.14, and 4.78 mm, respectively (Figure 1).

A total of 14 aortic dissections and 1 rupture were reported, all involving the thoracic aorta. The most common presenting symptoms were chest pain (75%) and

Figure 1. Average Aneurysm Growth From Baseline



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syncope (18%). Patients with aortic dissection or rupture had similar characteristics to the baseline cohort. Drugs used at the time of dissection or rupture included glucocorticoids (58%), methotrexate (8%),  $\beta$ -blockers (83%), aspirin (67%), statins (33%), and angiotensin-converting enzyme inhibitors (50%).

The mean maximal aneurysmal dilation at the time of dissection or rupture was  $54 \pm 11$  mm (range, 41-80 mm). The size of the aneurysm at the last follow-up was not predictive of dissection or rupture; intact aneurysms were a mean  $50 \pm 11$  mm ( $p=0.72$ ). There was no difference in aneurysm size at diagnosis between dissected or ruptured aneurysms ( $51 \pm 10$  mm) and those that did not dissect or rupture ( $49 \pm 11$  mm) ( $p=0.47$ ).

Emergent surgical repair was performed in 8 of 15 patients (57%) with dissection or rupture (88% in the ascending aorta). Medical management was attempted in 7 of 15 (47%), but 4 required surgical intervention. The overall mortality rate at 30 days was 14%.

There was an 8% incidence of acute aortic syndrome in this cohort of patients with GCA. The data analysis showed that dissection can occur at any size and that aortic size at diagnosis or follow-up did not predict dissection or rupture. This study demonstrated that acute aortic syndrome in patients with GCA has a significant risk for mortality.

## Higher Risk of Aortic Complications in GCA Patients With Positive PET

Written by Toni Rizzo

Aortic complications are an important cause of death in patients with giant-cell arteritis (GCA). According