Clinical Policy Bulletin: Canakinumab (Ilaris)

Number: 0881

Policy

I. Aetna considers canakinumab (Ilaris) medically necessary for the following indications:
A. Cryopyrin-associated periodic syndromes (CAPS) when the following criteria are met:

1. The member has genetic evidence of an CIAS1 (NLRP3) mutation based on DNA sequencing, and has classic signs and symptoms of familial cold autoinflammatory syndrome (FCAS) (recurrent, intermittent fever and rash that were often exacerbated by exposure to generalized cool ambient temperature [natural, artificial, or both]), or Muckle-Wells syndrome (MWS) (chronic fever and rash of waxing and waning intensity, sometimes exacerbated by exposure to generalized cool ambient temperature); and

2. The member is 4 years of age or older; and

3. There is clinical documentation of functional impairment resulting in limitations of activities of daily living; and

4. The member is not using a tumor necrosis factor blocking agent (e.g., adalimumab, etanercept and infliximab) or anakinra; and

5. The member does not have active or chronic infections including hepatitis B, hepatitis C, human immunodeficiency virus, or tuberculosis; and

6. The member is not receiving a live vaccine concurrently with canakinumab administration; or

B. Active systemic juvenile idiopathic arthritis (SJIA), for persons who have not responded to a trial of an NSAID alone, or whose initial symptoms include high fevers and painful polyarthritis (severe disease).

II. Aetna considers experimental and investigational the use of canakinumab in combination with a tumor necrosis factor blocking agent (e.g., adalimumab, etanercept, or infliximab) or anakinra.

III. Aetna considers canakinumab experimental and investigational for all other indications including the following (not an all-inclusive list) because its effectiveness for these indications has not been established:
Canakinumab (Ilaris) is a recombinant, human anti-human-IL-1beta monoclonal antibody. It is indicated for the treatment of CAPS, including FCAS and MWS in adults and children 4 years of age and older (Walsh, 2009).

The approval of canakinumab by the FDA in June 2009 was based on a 3-part, 48-week, double-blind, placebo-controlled, randomized withdrawal study of canakinumab in patients with CAPS (Lachmann et al, 2009). In part 1, 35 patients received 150 mg of canakinumab subcutaneously. Those with a complete response to treatment entered part 2 and were randomly assigned to receive either 150 mg of canakinumab or placebo every 8 weeks for up to 24 weeks. After the completion of part 2 or at the time of relapse, whichever occurred first, patients proceeded to part 3 and received at least 2 more doses of canakinumab. These investigators evaluated therapeutic responses using disease-activity scores and analysis of levels of CRP and SAA. In part 1 of the study, 34 of the 35 patients (97%) had a complete response to canakinumab. Of these patients, 31 entered part 2, and all 15 patients receiving canakinumab remained in remission. Disease flares occurred in 13 of the 16 patients (81%) receiving placebo (p < 0.001). At the end of part 2, median CRP and SAA values were normal (less than 10 mg/L for both measures) in patients receiving canakinumab; but were elevated in those
receiving placebo (p < 0.001 and p = 0.002, respectively). Of the 31 patients, 28 (90 %) completed part 3 in remission. In part 2, the incidence of suspected infections was greater in the canakinumab group than in the placebo group (p = 0.03). Two serious adverse events occurred during treatment with canakinumab: 1 case of urosepsis and an episode of vertigo. The authors concluded that treatment with subcutaneous canakinumab once every 8 weeks was associated with a rapid remission of symptoms in most patients with CAPS.

Dhimolea (2010) stated that canakinumab was approved by the FDA for the treatment of FCAS and MWS, which are inflammatory diseases related to cryopyrinCAPS. The drug is currently being evaluated for its potential in the treatment of chronic obstructive pulmonary disease, ocular diseases, rheumatoid arthritis, systemic-onset juvenile idiopathic arthritis, as well as type 1 and type 2 diabetes.

Sundy (2010) discussed approved and emerging drugs used to treat hyperuricemia or the clinical manifestations of gout. Results of several clinical trials provided new data on the safety and effectiveness of the approved urate-lowering drugs, allopurinol and febuxostat. New recommendations have been presented on appropriate dosing of colchicine for acute gout flares and potential toxicities of combining colchicine with medications such as clarithromycin. Emerging therapies, including pegloticase, the uricosuric agent RDEA596, and the IL-1 inhibitors, rilonacept and canakinumab, have shown promise in early and late phase clinical trials. The author concluded that recent publications demonstrate an opportunity to use existing gout therapies more effectively in order to improve both safety and effectiveness. Emerging therapies for gout show promise for unmet needs in selected gout populations.

Dinarello and colleagues (2012) noted that monotherapy blocking IL-1 activity in autoinflammatory syndromes results in a rapid and sustained reduction in disease
severity, including reversal of inflammation-mediated loss of sight, hearing and organ function. This approach can therefore be effective in treating common conditions such as post-myocardial infarction (MI) heart failure, and trials targeting a broad spectrum of new indications are underway. So far, 3 IL-1-targeted agents have been approved: (i) the IL-1 receptor antagonist anakinra, (ii) the soluble decoy receptor rilonacept, and (iii) the neutralizing monoclonal anti-IL-1β antibody canakinumab. In addition, a monoclonal antibody directed against the IL-1 receptor and a neutralizing anti-IL-1α antibody are in clinical trials.

Galeotti et al (2012) described the safety and effectiveness of IL-1-targeting drugs, anakinra and canakinumab, in patients with mevalonate kinase deficiency (MKD). A questionnaire was sent to French pediatric and adult rheumatologists to retrospectively collect information on disease activity before and after treatment with IL-1 antagonists from genetically confirmed MKD patients. The authors assessed the frequency of crises and their intensity using a 12-item clinical score built for the purpose of the study. A total of 11 patients were included. Anti-IL-1-targeting drugs were used continuously in all but 1 patient who received anakinra on demand. Daily anakinra (9 patients) or canakinumab injections every 4 to 8 weeks (6 patients, in 4 cases following anakinra therapy) were associated with complete remission in 4 cases and partial remission in 7. The median score during MKD attacks decreased from 7/12 before treatment to 3/12 after anakinra and 1/12 after canakinumab. The number of days with fever during attacks decreased from 5 before treatment to 3 after anakinra and 2 after canakinumab. Marked decrease of CRP and SAA protein were recorded. Side effects were mild or moderate; they consisted of local pain and inflammation at injection site, infections and hepatic cytolysis. The authors concluded that continuous IL-1 blockade brings substantial benefit to MKD patients. Moreover, they stated that controlled trials are needed to further evaluate the clinical benefit and treatment modalities in these patients.

Giampietro and Fautrel (2012) stated that IL-1β is emerging as a master mediator of adult-onset Still’s disease (AOSD) pathogenesis. This pleiotropic cytokine has a wide type of effects. As a key mediator of innate immunity, it is a potent pyrogen and facilitates neutrophilic proliferation and diapedesis into the inflamed tissues, which are key AOSD manifestations. The study of pro-inflammatory cytokines profiles in sera and pathological tissues of AOSD patients has shown elevated levels of IL-1β, these levels being highly correlated with disease activity and severity. These experimental evidences as well as the analogy with other auto-inflammatory diseases that share with AOSD clinical and biological characteristics have suggested the blockade of IL-1β as a possible new therapeutic option for the AOSD, especially in conventional therapy resistant cases. Anakinra, the first anti-IL-1 agent put on the market, has demonstrated capable to induce a rapid response sustained over time, especially in systemic forms, where anti-TNFα failed to control symptoms. While a growing number of evidences supports the utilization of anakinra in AOSD, a new generation of anti-IL1β antagonists is developing. Canakinumab and rilonacept could improve the management of this disease.

The American College of Rheumatology’s guidelines for management of gout (Khanna et al, 2012) noted that “Use of a biologic interleukin-1 (IL-1) inhibitor (anakinra 100 mg subcutaneously daily for 3 consecutive days; evidence B) or
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Kontzias and Efthimiou (2012) described the successful treatment of AOSD with canakinumab on patients refractory to anakinra and rilonacept. In many cases the expected positive therapeutic effect of short-acting IL-1 inhibitors is transient or completely absent, leading to the hypothesis that their short half-life may be associated with incomplete IL-1 blockade, given the cyclic nature of the disease. These investigators reported 2 cases of AOSD resistant to short-acting IL-1 blockade, which were subsequently treated with canakinumab. A retrospective chart review was conducted of patients diagnosed with AOSD in the authors’ regional referral center. Response to treatment was assessed by its effect on the systemic symptoms (resolution of fever and rash), polyarthritis (using the disease activity score 28 – CRP score), and the levels of serum ferritin. Canakinumab demonstrated sustained efficacy in both patients as evidenced by clinical and laboratory parameters with minimal adverse reactions. The authors concluded that this is the first documented report of successful use of canakinumab in AOSD patients refractory to traditional disease-modifying anti-rheumatic drugs and short- to moderate-acting IL-1 blockade. Moreover, they stated that prospective comparative studies are needed to validate canakinumab’s safety and effectiveness in the treatment of AOSD.

Lipsker and Lenormand (2012) stated that anecdotal observations suggested that IL-1 antagonists may be effective for the treatment of patients with different types of inflammatory dermatological diseases. These investigators reviewed the current evidence on the use of IL-1 antagonists in dermatology. A Medline search was performed combining the keywords: ”anakinra; canakinumab; rilonacept” and "skin; neutrophilic dermatoses; Sweet syndrome; pyoderma gangrenosum; hidradenitis suppurativa; Schnitzler syndrome; Still disease". The precise dermatological phenotype of patients with IL-1 antagonist-responsive auto-inflammatory disorders was analysed in order to compare it to related complex disorders. Double-blind randomized controlled trials have demonstrated the efficacy of these treatments in cryopyrinopathies with dermatological involvement including chronic infantile neurological cutaneous and articular (CINCA) syndrome, Muckle-Wells syndrome and familial cold urticaria. Anakinra is the only treatment for Schnitzler syndrome that is almost constantly efficacious, even in refractory disease, as attested by numerous case reports. It is also efficacious in the treatment of patients with adult-onset Still disease and systemic juvenile arthritis. Neutrophilic dermatoses constitute the cutaneous hallmark of IL-1-responsive auto-inflammatory disorders, and neutrophilic dermatoses could thus form an indication for this treatment. However, to-date, only 9 reports have been published showing efficiency in patients with Sweet syndrome, in 1 case of neutrophilic panniculitis, and in 2 cases of pustular psoriasis. Anakinra appears less efficacious in patients with pyoderma gangrenosum. The authors concluded that IL-1 antagonists are a first-line treatment in patients with Schnitzler syndrome and cryopyrinopathies. They could become important alternatives in patients with acute and febrile
neutrophilic dermatoses either unresponsive to or with contraindications to conventional treatments, but this requires confirmation by further clinical trials.

Ridker et al (2012) conducted a double-blind, multi-national phase IIb trial of 556 men and women with well-controlled diabetes mellitus and high cardiovascular risk who were randomly allocated to subcutaneous placebo or to subcutaneous canakinumab at doses of 5, 15, 50, or 150 mg monthly and followed over 4 months. Compared with placebo, canakinumab had modest but non-significant effects on the change in hemoglobin A1c, glucose, and insulin levels. No effects were seen for low-density lipoprotein cholesterol, high-density lipoprotein cholesterol, or non-high-density lipoprotein cholesterol, although triglyceride levels increased approximately 10 % in the 50-mg (p = 0.02) and 150-mg (p = 0.03) groups. By contrast, the median reductions in C-reactive protein at 4 months were 36.4 %, 53.0 %, 64.6 %, and 58.7 % for the 5-, 15-, 50-, and 150-mg canakinumab doses, respectively, compared with 4.7 % for placebo (all p values ≤ 0.02). Similarly, the median reductions in interleukin-6 at 4 months across the canakinumab dose range tested were 23.9 %, 32.5 %, 47.9 %, and 44.5 %, respectively, compared with 2.9 % for placebo (all p ≤ 0.008), and the median reductions in fibrinogen at 4 months were 4.9 %, 11.7 %, 18.5 %, and 14.8 %, respectively, compared with 0.4 % for placebo (all p values ≤ 0.0001). Effects were observed in women and men. Clinical adverse events were similar in the canakinumab and placebo groups. The authors concluded that canakinumab significantly reduced inflammation without major effect on low-density lipoprotein cholesterol or high-density lipoprotein cholesterol. They stated that these phase II trial data supported the use of canakinumab as a potential therapeutic method to test directly the inflammatory hypothesis of atherosclerosis.

Systemic juvenile idiopathic arthritis, previously referred to as Still’s disease or systemic onset juvenile rheumatoid arthritis, is a subset of juvenile idiopathic arthritis. Individuals with systemic juvenile idiopathic arthritis present with intermittent fever, rash, and arthritis. Children with this illness comprise between 10 and 20 percent of all cases of JIA. Children with systemic onset JIA require close supervision and careful monitoring as systemic complications, including drug reactions, macrophage activation syndrome, pericarditis, and other forms of internal organ involvement are more common in this subtype of JIA than in any other (Lehman, 2014).

Ruperto et al (2012) assessed the safety and effectiveness of canakinumab for the treatment of systemic JIA in 2 trials. In trial 1, these researchers randomly assigned patients, 2 to 19 years of age, with systemic JIA and active systemic features (fever; greater than or equal to 2 active joints; CRP, greater than 30 mg/L; and glucocorticoid dose, less than or equal to 1.0 mg/kg body weight/day), in a double-blind fashion, to a single subcutaneous dose of canakinumab (4 mg/kg) or placebo. The primary outcome, termed adapted JIA ACR 30 response, was defined as improvement of 30 % or more in at least 3 of the 6 core criteria for JIA, worsening of more than 30 % in no more than 1 of the criteria, and resolution of fever. In trial 2, after 32 weeks of open-label treatment with canakinumab, patients who had a response and underwent glucocorticoid tapering were randomly assigned to continued treatment with canakinumab or to placebo. The primary outcome was time to flare of systemic JIA. At day 15 in trial 1, more patients in the canakinumab group had an adapted JIA ACR 30 response (36 of 43 [84 %],
versus 4 of 41 [10 %] in the placebo group; p <0.001). In trial 2, among the 100 patients (of 177 in the open-label phase) who underwent randomization in the withdrawal phase, the risk of flare was lower among patients who continued to receive canakinumab than among those who were switched to placebo (74 % of patients in the canakinumab group had no flare, versus 25 % in the placebo group, according to Kaplan-Meier estimates; hazard ratio, 0.36; p = 0.003). The average glucocorticoid dose was reduced from 0.34 to 0.05 mg/kg/day, and glucocorticoids were discontinued in 42 of 128 patients (33 %). The macrophage activation syndrome occurred in 7 patients; infections were more frequent with canakinumab than with placebo. The authors concluded that these 2 phase III studies showed the efficacy of canakinumab in systemic JIA with active systemic features. The main drawback of the 2 studies was that patients without fever were excluded from participation. In a subset of patients with systemic JIA, systemic symptoms eventually resolve while chronic arthritis continues. Thus, the effectiveness of canakinumab in patients who have systemic JIA without fever cannot be deduced directly from these findings. Furthermore, information on the safety of canakinumab in patients with systemic JIA is limited, given the short duration of exposure to placebo in both trials and the use of a withdrawal design. The authors stated that longer-term safety data are needed.

Akgul et al (2013) performed a systematic review to analyze patients with familial Mediterranean fever (FMF), including juvenile patients who received treatment with biologics. A MEDLINE search, including articles published in English language between 1990 and May 2012, was performed. Patients who had Mediterranean fever variants but could not be classified as FMF according to Tel-Hashomer criteria were excluded. There is no controlled trial on the safety and effectiveness of biologics in FMF. A total of 59 (32 females and 27 males) patients with FMF who had been treated with biologics (infliximab, etanercept, adalimumab, anakinra, and canakinumab) were reported in 24 single reports and 7 case series. There were 16 children and 43 adults (7- to 68-year olds). Five patients were reported to have colchicine intolerance or had adverse events related to colchicine use, and the rest 54 were unresponsive to colchicine treatment. The authors concluded that the current data are limited to case reports, and it is difficult to obtain a quantitative evaluation of response to biologic treatments. However, on the basis of reported cases, biologic agents seem to be an alternative treatment for patients with FMF who are unresponsive or intolerant to colchicine therapy and seem to be safe. Moreover, they stated that controlled studies are needed to better evaluate the safety and effectiveness of biologics in the treatment of patients with FMF.

Horneff (2013) noted that the development of biologics has markedly changed the treatment of JIA, specifically that complete control of the disease and remission has today become the main goal of treatment, including preventing long-term damage and disability. The author’s review included an overview of the current treatment options using biologics in JIA. TNF inhibitors have emerged as the most commonly used biologics for the treatment of JIA. They were initially successful for the treatment of rheumatoid factor positive and negative polyarticular JIA, but have also been studied in patients with enthesitis-related arthritis, psoriatic arthritis, and extended oligoarthritis, and approval of at least etanercept is expected. Second-line biologics are abatacept and tocilizumab. For systemic onset JIA, tocilizumab, and the IL-1 inhibitors anakinra and canakinumab have been successfully studied and
in the treatment of JIA, biologics have emerged as potent drugs to control the
disease. The author further noted that new advancements will be crucial for
continued improvement in treatment options for JIA.

Moran et al (2013) examined if canakinumab or anakinra improved β-cell function
in recent-onset type 1 diabetes. These researchers performed 2 randomized,
placebo-controlled trials in 2 groups of patients with recent-onset type 1 diabetes
and mixed-meal-tolerance-test-stimulated C peptide of at least 0.2 nM. Patients in
the canakinumab trial were aged 6 to 45 years and those in the anakinra trial were
aged 18 to 35 years. Patients in the canakinumab trial were enrolled at 12 sites in
the USA and Canada and those in the anakinra trial were enrolled at 14 sites
across Europe. Participants were randomly assigned by computer-generated
blocked randomization to subcutaneous injection of either 2 mg/kg (maximum 300
mg) canakinumab or placebo monthly for 12 months or 100 mg anakinra or
placebo daily for 9 months. Participants and care-givers were masked to
treatment assignment. The primary end-point was baseline-adjusted 2-hr area
under curve C-peptide response to the mixed meal tolerance test at 12 months
(canakinumab trial) and 9 months (anakinra trial). Analyses were by intention to
treat. Patients were enrolled in the canakinumab trial between November 12,
2010, and April 11, 2011, and in the anakinra trial between January 26, 2009, and
May 25, 2011. A total of 69 patients were randomly assigned to canakinumab (n =
47) or placebo (n = 22) monthly for 12 months and 69 were randomly assigned to
anakinra (n = 35) or placebo (n = 34) daily for 9 months. No interim analyses were
done. A total of 45 canakinumab-treated and 21 placebo-treated patients in the
canakinumab trial and 25 anakinra-treated and 26 placebo-treated patients in the
anakinra trial were included in the primary analyses. The difference in C peptide
area under curve between the canakinumab and placebo groups at 12 months
was 0.01 nmol/L (95 % CI: -0.11 to 0.14; p = 0.86), and between the anakinra and
the placebo groups at 9 months was 0.02 nmol/L (-0.09 to 0.15; p = 0.71). The
number and severity of adverse events did not differ between groups in the
canakinumab trial. In the anakinra trial, patients in the anakinra group had
significantly higher grades of adverse events than the placebo group (p = 0.018),
which was mainly because of a higher number of injection site reactions in the
anakinra group. The authors concluded that canakinumab and anakinra were safe
but were not effective as single immunomodulatory drugs in recent-onset type 1
diabetes.

Otten et al (2013) conducted a systematic review of all available efficacy data from
11 randomized controlled trials performed in JIA with inclusion of biological
agents. If trials were comparable with regard to design and patients’
characteristics related to treatment outcome an indirect between-drug comparison
was conducted. On the basis of the equality of the trials, 6 trials were grouped into
two networks of evidence. Network 1, which included withdrawal trials evaluating
etanercept, adalimumab and abatacept in polyarticular course JIA, showed no
significant differences in short-term efficacy based on indirect comparisons.
Network 2 indirectly compared trials with parallel study design investigating
anakinra, tocilizumab and canakinumab in SJIA and found no differences in
comparative efficacy. The authors concluded that due to the small number of trials
and the observed differences between trials, no definite conclusions could be
drawn regarding the comparative effectiveness of the indirectly compared
biological agents. They recommended that comparability of future trials be
improved and noted that head-to-head trials are required to decide on the best biological treatment for JIA.

Russo et al (2013) conducted a single-center observational study to determine the short- and long-term efficacy and safety of 8-weekly canakinumab therapy in children with CAPS in routine clinical practice. Methods. Study participants were assessed every 8 weeks at a dedicated clinic and standardized assessments were the 10-domains DAS for CAPS, acute phase reactants (APRs), physician's global assessment of disease activity, Child Health Assessment Questionnaire (CHAQ) and Child Health Questionnaire Parent Form 28 (CHQPF-28). The primary endpoint of clinical improvement was defined as a reduction of DAS score 8 weeks after commencing therapy and secondary endpoints included sustained clinical improvement in APRs, relapses, CHAQ score and CHQPF-28 score. Results. Ten children with CAPS [eight Muckle-Wells syndrome (MWS), two chronic infantile cutaneous neurological articular (CINCA); median age 6.3 years] received 8-weekly canakinumab treatments at 2-8.7 mg/kg for a median of 21 months (range 12-31 months, with nine of 10 patients improving after the first dose: baseline median DAS of 7.5/20 decreased to 3.5/20 at 8 weeks (P = 0.04). This clinical improvement was sustained at a median follow-up of 21 months (range 12-31 months). It was noted that children with CINCA required higher doses of canakinumab than those with MWS. CHAQ and CHQ scores indicated improvement in functioning and health-related quality of life (HRQoL) and treatment was well tolerated, with no injection site reactions and no serious infections. The authors concluded that Canakinumab, although costly, is a safe and effective treatment for CAPS in children, leading to sustained improvement in disease activity, serological markers, functional ability and HRQoL.

Thompson et al (2013) noted that rupture or erosion of an unstable atherosclerotic plaque is the typical pathology and usual cause of acute coronary syndromes (ACS). Despite detailed understanding of the processes of lipid accumulation, thinning of the fibrous cap, and inflammation leading to plaque instability, there are no strategies in clinical use that uniquely target the unstable plaque. These investigators performed a critical review of recent publications on potential therapies that could be used to stabilize unstable plaque. They searched PubMed, other literature databases, drug development sites, and clinical trial registries to retrieve clinical studies on anti-inflammatory and lipid-modulating therapies that could be used to stabilize unstable atherosclerotic plaque. Multiple experimental targets involving lipid and inflammatory pathways have the potential to stabilize the plaque and expand the armamentarium against coronary artery disease. Randomized clinical trials of darapladib, methotrexate, canakinumab, and colchicine are well advanced to establish if plaque stabilization is feasible and effective in patients with ACS. The authors concluded that although there are still no agents in clinical use for plaque stabilization, there are important advances in understanding plaque instability and several encouraging approaches are being evaluated in phase III clinical trials.

Vanderschueren and Knockaert (2013) tested canakinumab in patients with Schnitzler syndrome. A patient with Schnitzler syndrome was treated with canakinumab, 150 mg subcutaneously injection every 8 weeks for 6 consecutive months. Injections were resumed in case of a flare following discontinuation. Canakinumab induced a swift and sustained clinical response,
with disappearance of fever and arthralgias, near abolishment of fatigue and rash, and substantial reduction of CRP levels. Interruption of canakinumab after four 8-weekly injections led to a flare 10 weeks after the last administration, which was countered as soon as canakinumab injections were resumed. The patient remained in complete remission. Canakinumab was well-tolerated. No injection site reactions, other adverse events, or laboratory abnormalities were observed. The authors concluded that canakinumab has potential for the treatment of Schnitzler syndrome.

Canakinumab received FDA approval for use in systemic juvenile idiopathic arthritis on May 9, 2014 (Novartis, 2013).

Dosing:

According to the FDA approved labeling, dosing for CAPS consists of 150 mg for CAPS patients with body weight greater than 40 kg and 2 mg/kg for CAPS patients with body weight greater than or equal to 15 kg and less than or equal to 40 kg. For children 15 to 40 kg with an inadequate response, the dose can be increased to 3 mg/kg. Administer subcutaneously every 8 weeks (FDA, 2013).

According to the FDA approved labeling, dosing for SJIA consists of 4 mg/kg (with a maximum of 300mg) for patients with a body weight greater than or equal to 7.5kg. Administer subcutaneously every 4 weeks (FDA, 2013).

CPT Codes / HCPCS Codes / ICD-9 Codes

Other CPT codes related to the CPB:

96372  Therapeutic, prophylactic, or diagnostic injection (specify substance or drug); subcutaneous or intramuscular

HCPCS codes covered if selection criteria are met:

J0638  Injection, canakinumab, 1 mg

ICD-9 codes covered if selection criteria are met:

277.31  Familial mediterranean fever [Muckle-Wells syndrome (MWS)]

708.2  Urticaria due to cold and heat [familial cold autoinflammatory syndrome (FCAS)]

714.30 - 714.33  Juvenile chronic polyarthritis [covered in patients aged 2 years and older]

ICD-9 codes not covered for indications listed in the CPB (not all inclusive):

249.00 - 250.03  Diabetes mellitus

273.1  Monoclonal paraproteinemia [Schnitzler syndrome]
274.00 - Gouty arthropathy
274.03

277.89 Other specified disorders of metabolism, other specified disorders of metabolism [Mevalonate kinase]

360.00 - Disorders of the eye and adnexa
379.9

411.1 Intermediate coronary syndrome

428.0 - 428.9 Heart failure

440.0 - 440.9 Atherosclerosis

490 - 496 Chronic obstructive pulmonary disease and allied conditions

702.8 Other specified dermatoses [inflammatory]

714.0 - 714.2, 714.4 - 714.9 Rheumatoid arthritis and other inflammatory arthropathies

714.4 - 714.9

The above policy is based on the following references:


