Relapsing polychondritis treatment guideline

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Relapsing polychondritis (RPC) is a autoimmune systemic disorder that is typically chronic and inflammatory in nature. The disease is characterized by the involvement of various tissues and organs, including the respiratory, cardiovascular, and auditory systems. The mainstay of treatment for RPC is anti-tumor necrosis factor-α (anti-TNF-α) therapy, which has shown efficacy in reducing symptoms and improving quality of life. However, the choice of therapy and the timing of treatment are critical factors to consider in managing this disease.

**Pharmacological treatment**

Treatment is empirically adapted step by step, based on the severity of involvement. The main purpose of treatment is to achieve symptom control and maintain respiratory stability and function. As the disease progresses, the treatment may need to be adjusted to manage the manifestations of the disease effectively.

**Disease activity index**

A disease activity index has been developed and validated to assess disease activity and treatment response. It includes parameters such as pain, swelling, and respiratory function. The index helps in monitoring the disease activity and response to treatment.

**Imaging studies**

Imaging studies, such as chest X-ray, CT, and MRI, are useful in assessing the extent and severity of involvement. Positron emission tomography (PET) scanning has also been shown to detect asymptomatic cartilaginous involvement. In addition, it may be more sensitive to early response to treatment.

**Respiratory involvement**

Respiratory involvement is a common manifestation of RPC, and the disease often presents with symptoms such as cough, dyspnea, and chest pain. The disease can lead to progressive respiratory compromise and can be life-threatening. The treatment of respiratory involvement is mainly supportive, and includes ventilation and oxygen therapy, cough suppressants, and anti-inflammatory agents.

**Cardiovascular involvement**

Cardiovascular involvement is a serious complication of RPC, and can be life-threatening. The disease can cause aortic valve regurgitation, aortic root dilatation, and left ventricular dysfunction. Early recognition and treatment are crucial to avoid progression to life-threatening complications.

**Renal involvement**

Renal involvement is a rare but serious complication of RPC. It can cause glomerulonephritis, interstitial nephritis, and chronic renal failure. Early recognition and treatment are critical to prevent permanent renal damage.

**Neurological involvement**

Neurological involvement is a less common but serious complication of RPC. It can cause cranial nerve palsies, peripheral neuropathy, and myelopathy. Early recognition and treatment are crucial to prevent permanent neurological damage.

**Ocular involvement**

Ocular involvement is a less common but serious complication of RPC. It can cause conjunctivitis, keratitis, and uveitis. Early recognition and treatment are critical to prevent permanent visual damage.

**Gastrointestinal involvement**

Gastrointestinal involvement is a less common but serious complication of RPC. It can cause esophageal dysmotility, gastroparesis, and enteropathy. Early recognition and treatment are critical to prevent permanent gastrointestinal damage.

**Musculoskeletal involvement**

Musculoskeletal involvement is a less common but serious complication of RPC. It can cause joint swelling, pain, and dysfunction. Early recognition and treatment are critical to prevent permanent musculoskeletal damage.

**Hematological involvement**

Hematological involvement is a less common but serious complication of RPC. It can cause anemia, thrombocytopenia, and leukopenia. Early recognition and treatment are critical to prevent permanent hematological damage.

**Other manifestations**

Other manifestations of RPC include respiratory tract involvement, cardiovascular involvement, and nervous system involvement. Early recognition and treatment are critical to prevent permanent complications.

**Conclusion**

Relapsing polychondritis is a complex and challenging disease, and the treatment is often tailored to the individual patient. Early recognition and treatment are crucial to prevent permanent complications and improve quality of life. Anti-TNF-α therapy is the mainstay of treatment, and the choice of therapy and timing of treatment are critical to consider in managing this disease.