Relapsing polychondritis (RPC) is a rare immune mediated disease which is associated with inflammation in cartilaginous tissue throughout the body. Especially the cartilaginous structures of ear, nose, joints and respiratory tract are affected. In around 30% of the cases an association with other diseases especially systemic vasculitis or myelodysplastic syndrome can be detected. The relative rarity of RPC has not permitted clinical trials to determine the efficacy and safety of therapy strategies. Often the medication in current use is largely empiric and based on case reports. Therefore different immunosuppressants such as cyclophosphamide, azathioprine, cyclosporine, mycophenolate mofetil and also new approaches like tumor necrosis factor alpha blockers (TNF-alpha antagonists) have been used for the treatment of severe manifestations of RPC with varying degrees of efficacy. This review gives a close look to clinical manifestation, diagnosis and also therapy options of RPC.