Title: Pulmonary manifestations in adult patients with chronic granulomatous disease

Ms. Helene 23232 Salvator hsalvator@yahoo.fr MD ¹, Prof. Dr Louis-Jean 23233 Couderc lj.couderc@hopital-foch.org MD ¹, Dr. Nizar 23234 Mahlaoui nizar.mahlaoui@nck.aphp.fr MD ², Dr. Elisabeth 23235 Rivaud e.rivaud@hopital-foch.org MD ¹, Dr. Colas 23236 Tcherakian c.tcherakian@hopital-foch.org MD ¹, Prof. Dr Olivier 23237 Hermine olivier.hermine@nck.aphp.fr MD ³, Prof. Dr Felipe 23238 Suarez felipe.suarez@nck.aphp.fr MD ³, Prof. Dr Olivier 23246 Lortholary olivier.lortholary@nck.aphp.fr MD ⁴ and Dr. Emilie 23247 Catherinot e.catherinot@hopital-foch.org MD ¹. ¹ Respiratory Diseases, Hopital Foch, Suresnes, France, 92150 ; ² CEREDIH, Hopital Necker, Paris, France, 75015 ; ³ Hematology, Hopital Necker, Paris, France, 75015 and ⁴ Infectious Diseases, Hopital Necker, Paris, France, 75015.

Body: Chronic granulomatous disease (CGD) is an inherited primary immunodeficiency (PI) characterized by severe infections and chronic inflammatory manifestations. Since most patients actually reach adulthood it is of importance to analyze pulmonary manifestations in adults. We retrospectively studied the whole medical records of 52 adult patients with CGD registered on the French National database for PI. Average age was 26 years (44 males / 8 females). CGD inheritance was X-linked in 36 patients and autosomal recessive in 11 patients. Median follow-up after the age of 16 was ten years. Before the age of 16, we registered 49 pulmonary infectious events (0.06 infections/year) and nine inflammatory events. After the age of 16, 45 respiratory infections occurred in 28 patients (0.2 infections/year): 23 invasive aspergillosis and 22 bacterial infections. Most of them occurred despite anti-infectious prophylaxis (34/40). We noted lack of fever or respiratory symptoms in 14/23 aspergillosis, diagnosed on systematic revue. For twelve patients, infectious diagnosis required pulmonary biopsy. We also registered 18 respiratory inflammatory events. Eight of them were contemporary of an infection (7/8 aspergillosis). We noted respiratory symptoms in 12 of these 18 events, requiring immunosuppressive drugs in ten cases (mostly corticosteroids or thalidomide).When they are performed, pulmonary biopsies mainly showed granuloma. CGD is no longer a pediatric disease. Lungs are frequently affected despite anti-infectious prophylaxis. Aspergillosis, often asymptomatic, is associated with disproportionate inflammatory reactions. Non infectious pneumonias are frequent and paradoxically require immunosuppressors.