

Introduction

Patients with Down Syndrome (DS) are known to be predisposed to middle ear infections and are more likely to develop complications. Hearing loss has been reported in nearly 80% of DS patients, as opposed to only 2.5% in otherwise healthy patients. Additionally, recurrent infections and resistance to aural antibiotics may lead to complicated infection, leading to the prolonged inflammation, increasing the risk of ear canal stenosis and cholesteatoma formation.

Management of chronic otitis media in patients with DS is often regarded as preference-sensitive care in that there is not one optimal intervention or treatment. Elucidating factors contributing to indications for imaging and referral to an appropriate subspecialty is of paramount concern to mitigate preventable hearing loss and the formation of progressive cholesteatoma. Our study aims to identify long-term sequelae and outcomes of COM in young adults with DS.

Methods

This single academic hospital-based retrospective study was both reviewed and approved by the institutional review boards of both the Tulane University School of Medicine (Ref #2022-375) and the Children’s Hospital of New Orleans, LCMC Health (#22-062), which granted a waiver of informed consent. Purposive sampling was conducted in June of 2023 to identify young adults (age of fifteen years+) with DS referred to neurotology for complaints of hearing loss associated with chronic otitis media during five years (January 2017 – January 2023). Excluded from this descriptive study were patients seen by neurotology for complaints unrelated to chronic otitis media and patients referred to otology under fifteen. Statistical analyses were performed using RStudio (3.6.0).

Results

Part One

Sixty-three patients with DS underwent evaluation for COM by pediatric otolaryngology. Of 126 ears, 117 were diagnosed with conductive hearing loss and six with mixed hearing loss (HL). Average time from first visit concerning for middle ear infection to diagnosis of HL was 6.5 months (Range: 0 to 42 months). HL resolution occurred in 4 ears with medical management and 23 ears with surgical intervention. The shorter the time from first visit to diagnosis of hearing loss was found to be predictive of HL resolution ($t(50)=1.95, p=0.05$). Time from first visit to intervention did not yield significant results in HL resolution.

Part Two

Five patients received a referral from pediatric otolaryngology to neurotology for complaints related to COM. Time from first attributed COM diagnosis to neurotology visit averaged 30 months (Range: -1 to 187). Three patients presented with otorrhea and hearing loss as secondary symptoms to COM, one presented with hearing loss and congestion, and one presented with only hearing loss. Imaging findings were consistent with cholesteatoma formation in three of the eight involved ears. All imaging showed evidence of infection. Post-intervention, four patients reported improvement of symptoms; however, complete resolution was not provided in all patients. One patient had worsening of hearing loss, warranting hearing aid intervention.

Figure 1: Diagnostic Timeline of Ear Pathology and Severity by Ear Laterality

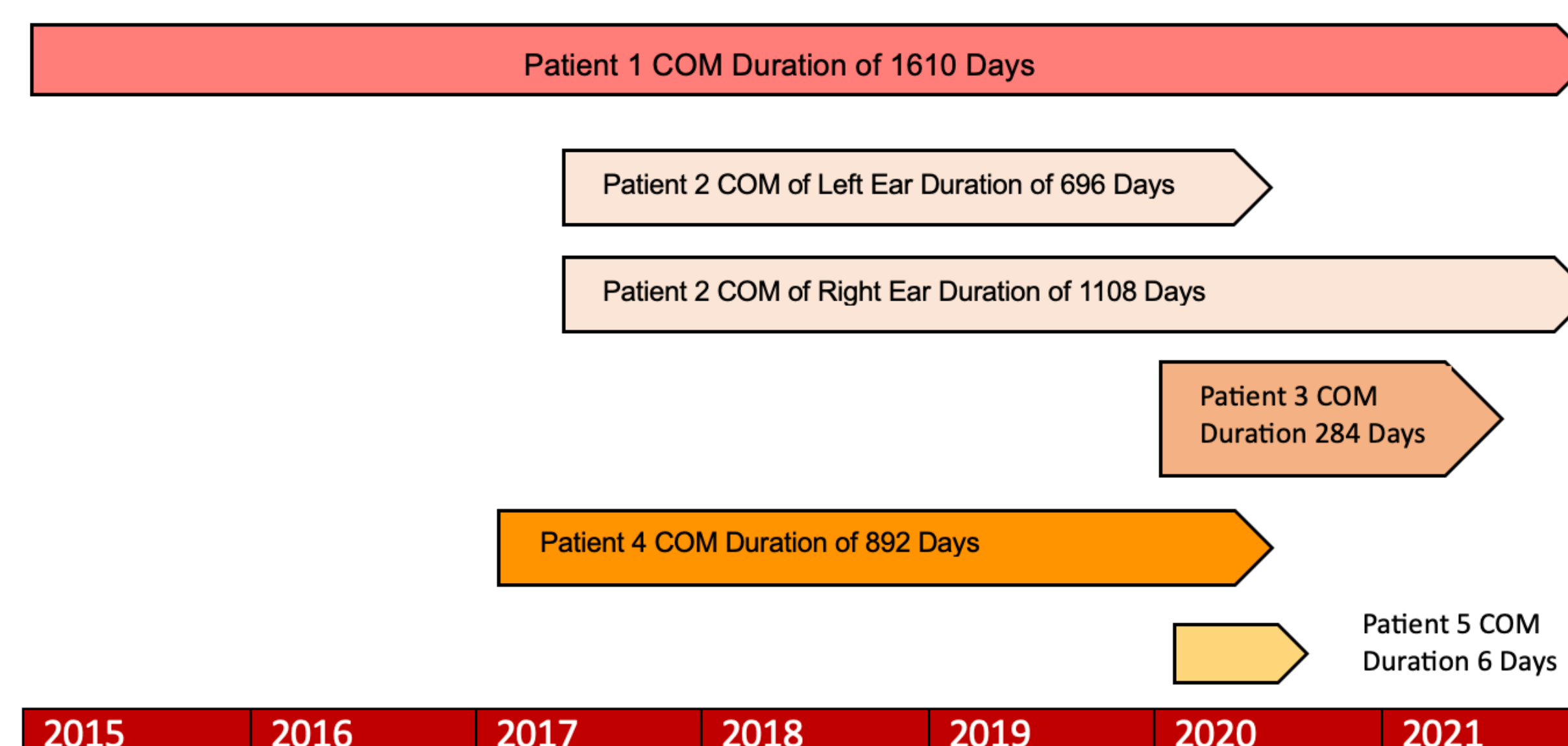
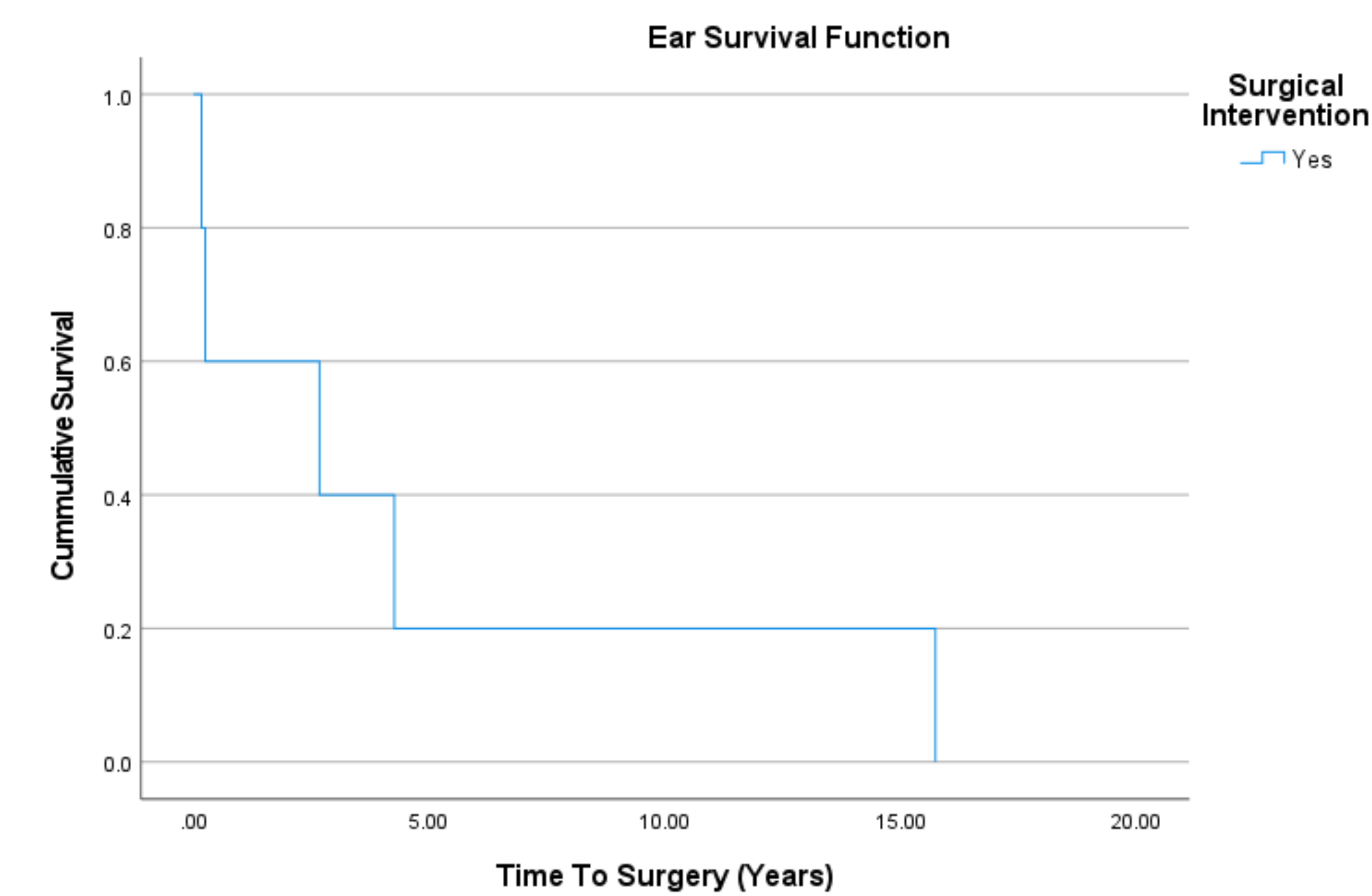


Figure 2: Ear Survival Curve as a Function of Surgical Intervention



Discussion

Our study demonstrates that increased surveillance and index of suspicion for severe complications of chronic otitis media, such as cholesteatoma, are warranted for patients with Down Syndrome. General practitioners and otolaryngologists should have a decreased threshold for imaging and referral to the appropriate subspecialty for patients with Down Syndrome that present with recurrent otitis media infections, have had multiple sets of tympanostomy tubes, and report no resolution of hearing loss symptoms. Further research on young adults and adults with Down Syndrome in otolaryngology is necessary, as life expectancy for these individuals has increased.

Citations

Austeng ME, Akre H, Øverland B, Abdelnoor M, Falkenberg ES, Kværner KJ. Otitis media with effusion in children with in Down syndrome. *Int J Pediatr Otorhinolaryngol.* 2013;77(8):1329-1332.

Tedeschi AS, Roizen NJ, Taylor HG, Murray G, Curtis CA, Parikh AS. The prevalence of congenital hearing loss in neonates with Down syndrome. *J Pediatr.* 2015;166(1):168-171.

Morris P. Chronic suppurative otitis media. *BMJ Clin Evid.* 2012;2012.

Hall A, Pryce H, Bruce IA, Callery P, Lakhanpaul M, Schilder AGM. A mixed-methods study of the management of hearing loss associated with otitis media with effusion in children with Down syndrome. *Clin Otolaryngol.* 2019;44(1):32-38.