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Review Article

Savant Syndrome: A Concise Review of the Causes, Symptoms, Diagnosis, and Treatment

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Abstract

Savant syndrome (SS) is a unique and rare condition in which individuals with developmental concerns, such as autism or central nervous system (CNS) diseases or illnesses, demonstrate exceptional 'islands of genius' that are strikingly juxtaposed to overall limitations.. These characteristics mean that these individuals exhibit special abilities in some areas. Special skills may often be in music, art, calendar calculations, arithmetic, or mechanical/visual/spatial capabilities. SS can be congenital (present from birth through infancy) or acquired following a brain injury or CNS dysfunction. Typically, it is associated with severe brain metabolic abnormalities, resulting in significant brain network alterations. These alterations are frequently explained by the 'tyranny of the left hemisphere' hypothesis, which states that the left hemisphere is suppressed for the right hemisphere to acquire savant abilities. Neuromodulatory procedures such as transcranial magnetic and direct current stimulation can briefly replicate these brain alterations. These approaches can help discover the potential of 'hidden talent by regulating the metabolism of the brain network. This review explores, summarizes, and discloses the historical and contemporary definition and understanding of SS and its association with autism spectrum disorder and other possible conditions.

Keywords: *Autism; Brain Metabolic Abnormalities; CNS Diseases; Neuromodulatory Procedures; Special Abilities*

Abbreviations

ABA: Applied Behavior Analysis; ASD: Autistic Spectrum Disorder; CAM: Complementary and Alternative Medicine; CBT: Cognitive Behavioral Therapy; CNS: Central Nervous System; CT: Computed Tomography; GTS: Gilles de la Tourette Syndrome; MRI: Magnetic Resonance Imaging; NAC: N-Acetylcysteine: rTMS: Repetitive Transcranial Magnetic Stimulation; SS: Savant Syndrome

Introduction

Individuals with savant syndrome (SS), despite having a severe mental or physical impairment, demonstrate "islands of genius": extraordinary abilities in music, mathematics, memory, or art. However, their abilities contrast starkly with their average intellect in other areas [1].

The 'islands of genius' associated with SS have a significant history. In 1783, *Gnothi Sauton*, a German journal of empirical psychology, published the first SS report, documenting the example of Jedediah Buxton [2]. Despite being unable to read or write his name, Mr. Buxton could perform quick mathematical operations such as multiplication or division of large numbers with high precision and speed compared to expert arithmetic [3]. In 1789, Rush, regarded as the founder of American psychiatry, provided an early account of Thomas Fuller's lightning-quick calculation skills. He said that Fuller' could understand virtually everything, both theoretical and practical, more complex than counting. For example, when asked how many seconds a guy who was 70 years, 17 days, and 12 hours old had lived, Fuller correctly responded in 90 seconds - 2,210,500,800, even accounting for the 17 leap years [4-6].

A century later, in 1887, in London, at the request of the Medical Society of London, Dr. Langdon Down, the director of the Earlswood Hospital, delivered the illustrious Lettsomian Lecture. The first explicit description of SS was given in the lecture. He identified 10 individuals with extraordinary skills and talents set against significant limitations. For example, one of his patients could recite "the rise and fall" of the Roman Empire forward and backward from memory. While some children drew brilliantly, others had a comparative blank in all other thinking capacities. However, other young people demonstrated musical talent, mathematical brilliance, and precise timekeeping skills, all combined to form a clinical picture of SS [1,2,7].

Édouard Seguin coined the phrase "idiot savant" in 1870, and George W. Grabham attributed it to the *British Medical Journal* in 1875. Before Binet's recommendation in 1905, 'idiot' was often used as a technical term; however, the phrase was used for SS for a considerable period, even after Binet suggested its discontinuation [7].

In 1914, Tredgold, an employee of Earlswood Hospital, published a detailed account of SS in a chapter of his well-known textbook, *Mental Deficiency*. This landmark chapter, which was continued in subsequent editions for many years, described approximately 20 more examples from various physicians [2]. Down's work and Tredgold's illustrations (in his 1914 book, *Mental Deficiency*) provide powerful representations of savants' abilities. Indeed, Tredgold's categorization of skill domains was astonishingly foresighted: the areas he emphasized remain the most commonly documented in the current scientific literature [9].

Several notable individuals were identified as 'idiot-savants'. Gottfried Mind (1768–1814) earned the nickname 'The Cat's Raphael for his lifelike portrayals of animals. Oliver Sacks recounts about twin brothers who could not read but promptly detected the number of matches dropped from a box and swapped prime numbers. Nadia, at the age of 4 years, created stunning doodles comparable to Raphael's. Other impressive contemporary savants include Stephen Wiltshire, an autistic artist whose astonishingly precise and detailed landscape paintings have an incredible quality and output. For instance, after a quick helicopter trip above Tokyo, he produced a 360-degree cityscape without needing memory aids. In his book In the Key of Genius, Adam Ockelford, a British composer and musicologist, writes about the abilities of his student with SS, Derek Paravicini as a pianist. He recalls Derek's interest in music and early progress in this field despite blindness and intellectual difficulties. The savant phenomenon was made more widely known by Darold Treffert's seminal book

Extraordinary People, which included several case studies and his hypotheses about the processes that account for the manifestation of savant skill [9].

Although SS is a fascinating phenomenon in studying human differences and cognitive psychology, several myths and misconceptions about such individuals persist (Figure 1) [2,5,6,10].

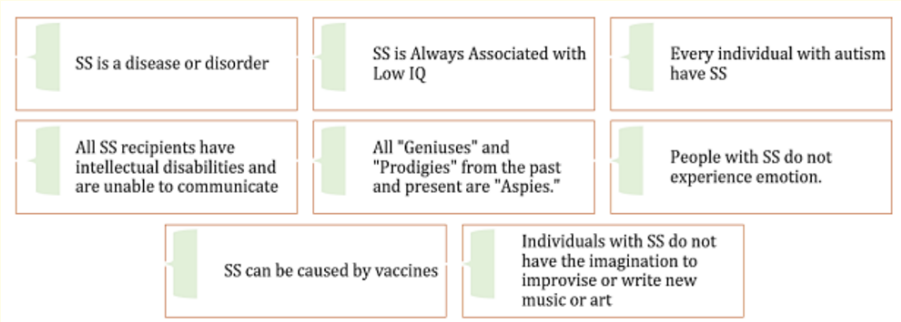

Figure 1: *Myths and misconceptions about savant syndrome [2,5,6,10].*

Discussion

Savant syndrome

SS is a rare and exceptional disorder in which individuals with autism, other severe mental impairments, or profound mental illnesses demonstrate extraordinary islands of competence or brilliance that stand out in stark contrast to their overall disability (or a person's overall character) [8,11]. The disorder can be congenital (existing from birth) or acquired (can arise following brain injury) [1]. Several examples of the congenital type can be quoted, including calendar calculators who can quickly determine the day of the week for any previous date; musical geniuses with perfect pitch, and hyperlexics, who, in one case, can read a page in 8s and remember the material with a recall rate of 99.9%. Other artistic abilities and talents, such as 3D sketching, memory for maps, poetry, painting, and sculpture, have also been reported. For example, a savant could memorize the Pi value up to 22,514 places without an error [7].

Seth F. Henriett, commonly known as "Rain Girl," is the most well-known SS autistic artist, poet, painter, and writer from Hungary. She is known to have inherited savant talents from her mother. However, as a youngster, she had received a false diagnosis of mental disability. Although intelligent, her understanding of numbers is not fully established, with a probable diagnosis of dyscalculia. Additionally, she struggles with communication despite exceptional verbal and visual skills [8].

Acquired SS develops in previously neurotypical individuals with a head trauma, stroke, dementia, or other CNS incidents (post-traumatic). Savant skills may suddenly manifest to a prodigious degree in such individuals [12]. For example, at 11 years of age, Orlando Serell lost consciousness and later complained of a terrible headache after being struck on the left side of his skull by a ball. After his headache subsided, Serell's memory improved dramatically, and his ability to perform accurate mathematical calculations [13]. Sudden SS, in contrast, is a newly recognized disorder in which remarkable talents emerge unexpectedly without an apparent underlying disability or brain damage. Sudden SS is distinct from a propensity to learn new skills (Figure A) [12].

Sudden Savant

- ✓ The skill manifests itself suddenly, with no prior interest or talent in the newly acquired ability.
- ✓ Neither CNS damage nor a disease or evident inciting event exists.
- ✓ The new abilities are combined with a thorough understanding of the fundamental principles governing music, art, or arithmetic.
- ✓ The obsessive-compulsive aspect of the skill is present at first; there is an overwhelming want to play music, create art, or perform calculations.

As with both congenital and acquired SS, it is a cause as well as a gift. [12]

Figure A

SS signs and symptoms

Signs and symptoms of SS can be recognized from childhood. The patient experiences both the loss of capabilities and the aberrant ability to perform something exceptional at different times (spatial skills, music, calendar and numerical calculations, art, mechanical abilities, and good memory). Figure 2 details the signs and symptoms of SS [14-17].

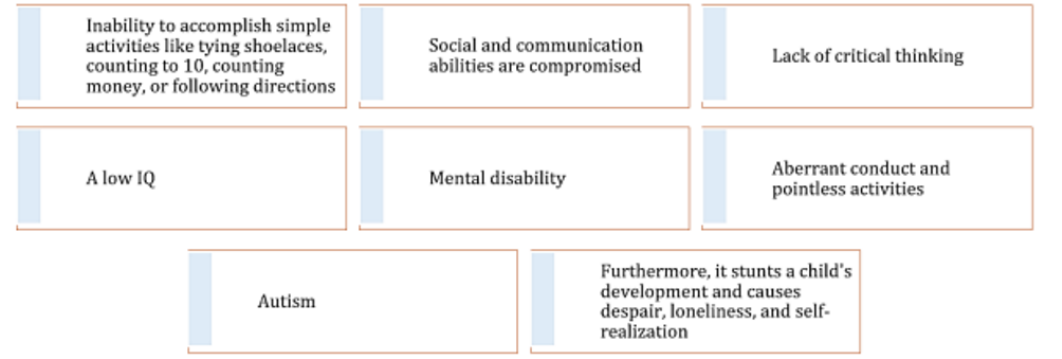

Figure 2: *Signs and symptoms of savant syndrome [14-17].*

SS characteristics

The condition is described as both a blessing and a burden. The ability has reportedly been known to manifest unexpectedly and mysteriously and vanish just as quickly. These specialized abilities can be grouped into 3 main categories: prodigious savants, talented savants, and splinter skills (Figure 3) [1,2,5,11].

SS mechanisms

Psychological: Generally accepted cognitive theories have yet to fully explain the combination of skills and impairment in savants [18]. Individuals with or without autism have been proposed to be susceptible to detail-focused processing [9]. Another explanation is that

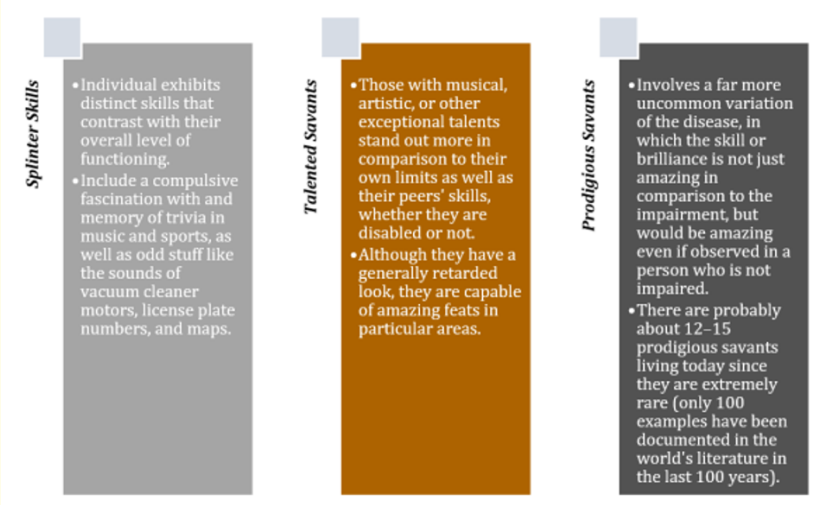

Figure 3: *Characteristics of savant syndrome [1,2,5,11].*

savants hyper-systematize, which gives the appearance of brilliance. The empathizing-systemizing hypothesis categorizes people based on their propensity to empathize with others instead of systematizing information about the outside world. Hyper-systemizing is just an extreme stage in this theory [19]. Also, their keen attention to detail is due to their unusually enhanced sensory perception (or hypersensitivity) [19,20]. Certain savants have also been proposed to work by directly accessing deep, unprocessed knowledge that resides in all human minds but is not ordinarily accessible to conscious awareness [21].

Neurological: SS occurs in individuals with developmental diseases such as Tourette syndrome and Asperger syndrome; individuals with CNS abnormalities such as injury to the left anterior temporal lobe, a region of the brain that is important for processing sensory information, identifying objects, and establishing visual memory, and individuals with front temporal dementia. These are often right-hemispheric skills (i.e. non-symbolic, tangible, intuitive, spatial-visual construction activities), contrary to left-hemispheric skills, which are more sequential, logical, and symbolic, including language specialization. When the left side of the brain is damaged, the right compensates, releasing additional potential.

According to Treffert., *et al.* (2009), an injury to the left hemisphere releases the right brain from the 'tyranny of the left hemisphere'. It allows the right brain to compensate for the damage, developing prodigious abilities [2]. One genius demonstrated an absence of corpus callosum (which integrates the motor, sensory and cognitive fibers between the two sides of the brain). In their model of autism, Rubenstein and Merzenich (2003) proposed that SS may be generated by an elevated ratio of excitation-to-inhibition in sensory, mnemonic, social, and emotional systems. They also highlighted the effects of genetic and environmental factors on the neurological system responsible for this higher ratio [22].

Incidence and prevalence of SS

SS appears to affect one in a million people [23]. However, comprehensive epidemiological investigations of savant abilities still need to be improved. Current prevalence estimates are based on surveys of parents or caregivers, frequently using chosen samples. In the late

1970s, from a postal survey of 5401 parents of children with autism, Bernard Rimland reported examples of 531 (9.8%) claimed savant talents. The most prevalent skills were music (53%), memory (40%), math/calculation (25%), and painting (10%), with 53% claiming more than a single unique ability. Bolte and Poustka, in a more recent survey, reported that 33 (13%) of 254 individuals with autism have a specific skill. This survey used standardized diagnostic interviews with parents [9].

A 2015 research project—that examined information from a registry of 319 individuals with SS—also discovered sex disparities, with 79% of their sample being men and 21% women [24]. Compared to autistic disorder, which has a male-to-female ratio of approximately 4:1, SS has a male-to-female ratio of roughly 6: 1 [10].

SS diagnosis

Approved diagnostic criteria need to be improved for SS, making it impossible for a physician to diagnose the condition [25]. Moreover, SS is not recognized in the medical/clinical diagnostic system, the International Classification of Diseases, or the Diagnostic and Statistical Manual of Mental Disorders [8]. However, the Sussex Savant Questionnaire has recently been developed to characterize the condition. The questionnaire determines the presence and type of an individual's ability in several categories [25].

SS association with other conditions and disorders

The remarkable islands of mental competence among otherwise mentally disabled individuals define SS. Interestingly, there is a significant degree of concordance between SS and autistic spectrum disorder (ASD), with 10 - 30% of patients with ASD demonstrating extraordinary talents and around 50% of patients with SS also having ASD [2,26-30]. In general, mutations in the PTEN gene, fragile X syndrome (*FXS*), or neurofibromatosis (*NF1*) promote the over-activation of the PI3K/Akt/mTOR pathway, resulting in autism-like behaviors [31]. According to the SSM Health Treffert Center, about 1 in 10 people with autism have SS [25].

The high similarity between ASD and SS may be attributed to a shared etiology or a mechanism by which one disease predisposes individuals to develop the other [32]. Nurmi., *et al.* (2003) claimed that chromosome 15q11-q13 is genetically associated with autism and SS [33], but Ma., *et al.* (2005) were unable to confirm this conclusion [34]. Despite the deliberations on the genes involved in SS, there is little reason to dispute that the disease has a genetic etiology.

In a Scandinavian sample, Steffenburg., *et al.* (1989) discovered that identical twins had an autism concordance rate of 90% compared to fraternal twins, who had 0% [35]. In a British sample, Bailey., *et al.* (1995) found that identical twins had a causal association of 73% compared with fraternal twins, who had 0% [36]. From several family and twin studies, Bailey., *et al.* (1996) found the heritability of autism to be at least 90% [37]. Therefore, considering the co-occurrence of autism and SS, it is highly plausible that SS also has a genetic etiology.

According to the Enhanced Perceptual Functioning paradigm of Mottron., *et al.* (2009), the high incidence of unique talents in people with ASD may be due to overactive brain areas involved in perceptual processes [20]. Veridical mapping has been further proposed as a specific mechanism that accounts for the role of perception in the manifestation of exceptional skills among people with ASD [32].

Similarly, in their 'Fractionable Triad Proposal', Happé and Vital stated that more confined and repetitive behaviors and interests, compared to slightly reduced social and communication difficulties, were the primary causes of the high prevalence of ASD-like symptoms in savant-autistic children. The autistic trait of a detail-orientated cognitive style (poor coherence) predisposes the person to be a genius, and the existence of savant talents may help in social competence [38].

Young (1995) conducted one of the most exhaustive investigations. He used standardized testing methods to obtain data from 51 individuals with SS. All the study patients demonstrated autistic traits. Of the 51 individuals, 41 met the diagnostic criteria for autism, while

the remainder also had abnormalities in their mental functioning. Regarding aptitude, 12 were prodigious savants, 20 were considered gifted, and 19 had 'splinter' qualities. The data below details the typical traits of savants, as Young identified (Figure B) [39].

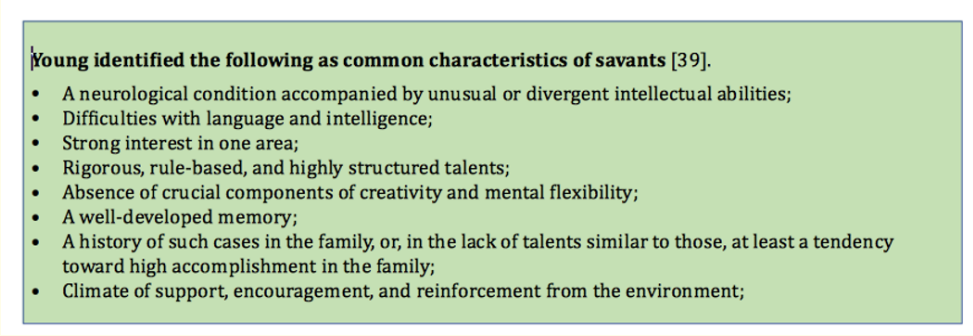

Figure B

However, SS is not always associated with an autistic condition. In institutionalized patients diagnosed with mental retardation, Hill (1977) discovered that the prevalence of SS was 0.06% (1 per 2000 patients) [40]. In a survey of 583 institutions, Saloviita., *et al.* (2006) discovered a prevalence rate of 1.4 per 1000 patients [41]. Irrespective of the actual numbers, mental retardation and other developmental disabilities are more prevalent than autistic disorder; therefore, it is acceptable to assume that around 50% of people with SS have autism as the underlying problem and the other 50% have another kind of developmental disability, mental retardation, or a CNS injury or disease. Publications on acquired SS and epilepsy in the global literature are also scant. Oliver Sachs reported a case of a person with 'high temperature, weight loss, delirium, and maybe convulsions.' After being sick, the individual started painting incredibly explicit scenes that his co-workers thought he could not have created before the sickness [7].

Neurodevelopmental disability "Gilles de la Tourette syndrome" (GTS)—which causes a range of tics, echolalia, and coprolalia—is another condition occasionally cited in the literature on savants. The prevalence of autism in GTS is significantly higher than predicted based only on population base rates. Therefore, it is not unexpected that many savant case studies report the existence of GTS, sometimes without a diagnosis of autism but with the common characteristics that may be crucial for developing savant skills (such as obsessive-compulsive behaviors/tendencies) [42].

SS affects particular categories of abilities

When considering the whole range of human abilities, savant abilities are limited to music, art, calendar calculations, lightning-fast calculations, and mechanical or spatial capabilities (language or memorization) (Figure 4). Moreover, these specialized skills are usually related to the right-hemispheric function [10].

Other abilities, such as extraordinary sensory discernment in smell, touch, or vision, including synesthesia; flawless understanding of time passing without a clock; and exceptional expertise in certain subjects, such as neurophysiology, statistics, or navigation, are also seldom documented. Often, there is more than one talent. Any unique ability is always complemented by extraordinary memory. Some observers see memory as a distinct particular skill [10]. Figure 5 lists some of the famous and notable cases of SS [50-52].

Figure 4: *Savant skills usually occur in particular categories of abilities [8,18,43-49].*

Figure 5: *Famous and notable cases of savant syndrome [50-52].*

Induction of SS

The extraordinary abilities of savants are proposed to be latent in everyone but are not easily accessible without a rare type of brain dysfunction. However, there are now several reports of intentionally generated savant-like abilities (including sketching, proofreading, numerosity, and false memory reduction) by altering the power of the left frontotemporal lobe to operate [21].

According to specific theories, savants have unique access to sensory input that needs to be integrated into a complete picture, which accounts for their outstanding talents. Everyone is assumed to have this more superficial and literal knowledge [53,54]. Yet, it is generally inaccessible unless there is a rare kind of brain damage, which can only be achieved artificially [55,56]. Snyder., *et al.* (2003) sought to mimic such brain damage in 11 healthy individuals by applying low-frequency magnetic pulses to the left frontotemporal lobe for 15 min. Participants were given a minute to recall a dog, horse, or face before, during, immediately after, and 45 minutes after receiving repetitive transcranial magnetic stimulation (rTMS) treatment [55].

The findings showed that the low-frequency rTMS of LATL did not systematically improve artistic performance. However, it significantly altered the drawing scheme or procedure for 4 of the 11 individuals. Some of those who were 'facilitated' also showed improved proofreading skills. These modifications can be attributed to the inhibitory effect of low-frequency rTMS. They result from disabling rather than stimulating a portion of the brain.

Alterations in perception or electroencephalogram-assisted feedback, brain damage, and magnetic stimulation can potentially enable savant-like abilities [54,57]. In his book, *The Mind's Eye*, Sacks, a neurologist, states that he could create drawings as exact as a camera only after using amphetamines. Mescaline-induced moods are associated with early (savant-like) cave art. Interestingly, after magnetic stimulation, 3 of our 4 positive responders reported experiencing changed psychological states [58,59].

SS treatment approach

Surprisingly, SS is neither an illness nor a disorder and, therefore, cannot be cured. It is a situation in which a person with a fundamental brain disorder shows exceptional memory and skills [60]. In 1930, Philips opined that developing potential is considerably more advantageous than dealing with specialized abilities, and the 'fault' can be improved. Whatever the specific capability, such as music, painting, or arithmetic, supporting and nurturing it eventually results in increased linguistic, social, and daily living abilities without sacrificing or losing those special skills as progress is made in other domains of functioning and learning. Instead of perceiving these unique talents as frivolous, special skills can be used to channel these talents more effectively [10]. For prodigious savants, early identification, as with any other disability, and deliberate encouragement are significant variables to improve skills. Assisting savants in attaining a higher level of general functioning may sometimes result in losing their specific abilities. Although data supporting this notion are scarce, developing skills may help improve socialization, communication, and self-esteem [43].

Patients with SS are often seen by developmental pediatricians, pediatric neurologists, psychologists, or psychiatrists.

Individuals with SS-related talents may feel different from their peers, causing social isolation and other concerns. These concerns can lead to depression and other psychosomatic conditions. Therefore, individuals with SS may receive treatment similar to that for autism. However, this therapy regimen may involve various strategies depending on the symptoms' nature and degree of intensity [25].

- **Cognitive behavioral therapy (CBT):** CBT is a short-term systematic psychotherapy focusing primarily on the present and helping to change problematic thinking patterns and behaviors. CBT is based on the perception of life events, and the thought-frame process affects behaviors and feelings. The main goal of therapy sessions is to explore and create strategies for coping with daily problems and behaviors. CBT has successfully treated bipolar disorder, anxiety, and other mental illnesses [61,62].

- **Educational interventions**: Applied behavior analysis (ABA), developmental models, organized instruction, speech-language therapy, social skills therapy, occupational therapy, and CBT are examples of educational methods that are often used. Interventions use one of these methods to fully address autistic traits or concentrate treatment on a specific area of deficit [63,64]. For example, early and rigorous ABA therapy improves communication and adaptive functioning among preschoolers. It is also well established to enhance your intellectual achievements [65].
- **Medications**: Medications are considered when behavioral therapy does not seem to be effective. Risperidone and aripiprazole are atypical antipsychotic drugs approved by the US Food and Drug Administration for treating self-destructive and violent behaviors. However, it is vital to consider its risks versus benefits due to the potential of people with autism to respond to these medications differently. For example, children with comorbid inattentiveness or hyperactivity may benefit from the stimulant drug methylphenidate. Simultaneously, selective serotonin reuptake inhibitor antidepressants, such as fluoxetine and fluvoxamine, are known to reduce repetitive and ritualistic behaviors [63].
- **Complementary and alternative medicine (CAM)**: It helps to address the behavioral symptoms associated with ASD by addressing biochemical or physiological conditions [66]. Enhanced oxidative stress leads to protein, DNA, and cell damage. N-acetylcysteine (NAC) and methylcobalamin (B12) help reduce oxidative stress. A 12-week randomized placebo-controlled study found that NAC helped improve irritability in Young adults with autism [67].
- **Social and speech training:** Reports on the benefits of social skills training in people with autism are scant. In 2000, Nakamura and colleagues analyzed the case of a person with SS and Asperger syndrome. They found that training in social skills, instruction in basic mannerisms, and encouragement using a therapy companion are effective management options.
- **Creative activities:** For individuals with limited or non-existent speech (or word knowledge), art therapy can be offered as an alternative non-verbal form of communication. In addition, these strategies support self-identity, which can easily be lost in a substantial mental facility of people with mental disabilities [68].

Future directions

Since the initial description of SS by Down in 1887, advances have been made in comprehending this exceptional condition. Although several concerns persist, there is a growing interest in resolving them. This intention may be attributed to the rapid development of modern technologies that allow the study of functions, rather than just the anatomy, of the brain [10].

The biological foundation for the illness and the precise mechanism(s) that produce the extreme inequalities between regions of prodigious skill and otherwise modest intellectual capacities are being investigated. Studies that combine examinations of brain architecture, connection, and function will be highly advantageous. SS, similar to autism, is a multifaceted condition, and no single theory can account for all manifestations [10].

Modern investigative technologies help study brain architecture. The surface and deep architecture of the brain can be visualized using remarkably high-resolution computed tomography (CT) and magnetic resonance imaging (MRI), allowing for an in-depth examination of the brain's anatomy. Brain function studies, such as positron emission tomography, single-photon emission computed tomography (SPECT), or functional MRI, reveal information about the brain at work rather than simply observing the brain architecture, thus providing considerably more information about SS. Diffusion tensor imaging, an even more contemporary imaging technology, offers visual pictures of the connection between the brain's hemispheres. It has been beneficial for studying performance talents in the arts and music.

The intersection of prodigies, savants, and genius is significant and, in some respects, relatively narrow. Therefore, this interdisciplinary, multimodal, and compare and contrast research must include such individuals. Such investigations can clarify the argument between general and distinct intelligence. Some academics contend that savants offer a distinctive perspective on the creative process. Essential

details on how the brain works, how it changes, how the CNS is compensated, how to recruit brain cells, and how it heals have been previously revealed through earlier research [2,5].

Disability education in public schools improves acceptance and reduces negative stereotypes associated with disabilities. Integrating them into society makes kids less likely to be stigmatized as odd and unappreciated contributors. As a result, typical peers will be exposed to disabilities more frequently and grow in understanding and acceptance.

A skill should not be silenced by mainstream schooling; it must instead be emphasized. Due to liability concerns and the perception that all people with disabilities lack the necessary intellect for a particular job, employers are reluctant to recruit differently-abled individuals. However, the organization thrives due to the high caliber of work that SS individuals create if employers know the various traits associated with SS, such as organization, focus, and intense general work habits [69].

The intersection of prodigies, savants, and genius is significant and, in some respects, relatively narrow. Therefore, interdisciplinary, multimodal, compare and contrast investigations should include such individuals. Such studies can clarify the contention between general and distinct intelligence. Moreover, savants offer a distinctive perspective on the creative process itself.

Conclusion

Although SS is a rare disease, it has been discussed in the psychological and medical research literature for over a century. Individuals with SS demonstrate exceptional 'islands of mental capacity' despite having mental impairments. This review has discussed experimental results and theoretical explanations of SS from published literature. It has also covered evidence of the close association of SS with autism, considering savant skills in individuals with other clinical conditions. However, SS is now as mysterious as it was initially described. Research efforts are expected to continue uninterrupted considering the numerous unsolved concerns regarding the presence and framework of savant talent.

Conflict of Interest Statement

The authors declare that this paper was written without any commercial or financial relationship that could be construed as a potential conflict of interest.

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