

A Review of the Savant Syndrome and its Possible Relationship to Epilepsy

by

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Abstract

The goal of this paper is to review the Savant Syndrome (SS), characterized by outstanding islands of mental ability in otherwise handicapped individuals. Two forms exist: the congenital and acquired form. Among the many examples of the congenital form are the calendar calculators, who can quickly provide the day of the week for any date in the past. Other examples are the musical savants with perfect pitch and the hyperlexics, who (in one case) can read a page in 8 seconds and recall the text later at a 99% level. Other types of talents and artistic skills can be found, involving 3-D drawing, map memory, poetry, painting, sculpturing, including one savant who could recite without error the value of Pi to 22,514 places. The acquired form refers to the development of outstanding skills after some brain injury or disease, usually involving the left fronto-temporal area. This type of injury seems to inhibit the 'tyranny of the left hemisphere', allowing the right hemisphere to develop the savant skills. One other way to inhibit the left fronto-temporal area is to use transcranial magnetic stimulation in normal subjects and nearly one-half of these subjects can then perform new skills during the stimulation that they could not perform before. This type of finding indicates the potentiality in all of us for the development of savant skills under special circumstances. Explanations of the congenital SS include enhanced local connectivity as a compensation for underconnectivity of long-range fibers, but also weak central coherence, replaced by great attention to details, enhanced perceptual functioning and obsessive pre-occupation with specific interests.

I. Introduction

The Savant Syndrome (SS) is characterized by remarkable islands of mental ability in otherwise mentally handicapped individuals [1]. Treffert and Wallace [2] have traced the history of the SS, pointing out that in 1789 Dr. Benjamin Rush, often considered the Father of American Psychiatry, described the outstanding mathematical ability of one man, named Thomas Fuller. Nearly 100 years later in 1887 Dr. J. Langdon Down, well known for the syndrome with his own name, and supervisor of the Earlswood Asylum in London, referred to 10 patients with the SS as “idiot savants”. An even earlier reference was added by Foerstl [3] who discussed a man named Jedediah Buxton, described in February 1751 in the magazine, “Gentleman”. Mr. Buxton could not write his own name but could do quick mathematical calculations, like multiplying a 39 digit number by itself.

The goal of this study is to describe the SS and also to investigate the possible relationship of this syndrome to epilepsy, especially because the SS are often found in autistic patients who often have seizures.

2. Method

The Medline Section on the Internet provided references to published studies on the “Savant Syndrome”.

3. Different forms

3.1 Congenital SS

Treffert [1] has indicated that nearly one-half of those with the SS are individuals with autism (A) and the other one-half have some other type of developmental disability.

3.1.1 Calendar calculators (CC)

One of the most common forms of the SS are the CC. These individuals can readily identify the day of the week for any given date in the past. Different

authors have reported various characteristics of the groups that they have studied. Dubischar-Krivec et al.. [4] reported shorter reaction times and fewer errors for present or past dates than controls, but no differences from controls were found for future dates. Thioux et al.. [5] described Donny, a young autistic savant, “who is possibly the fastest and most accurate calendar prodigy ever described”. The title of this report likely justifies the latter statement : “The day of the week when you were born in 700 msec”. O’Connor and Hermelin [6] reported that calculating speeds were similar in 10 year old savants as in adult savants. Performance could not be accounted for by practice alone and no improvement was detectable over time.

Other authors have pointed out various normal characteristics in this group. An example is that savants did not differ from controls for short – or – long term memory. However, the savants showed a recall superiority for long-term retention of calendrical material [7]. Others [8] have described in one savant poor explicit knowledge of calendar structure and also that error rate and response latency increased with temporal remoteness of dates under consideration. Mottron et al. [9] also dealt with errors, reporting that testing of all the dates in a year revealed a random distribution of errors which were not stable across time. Also, that particular savant was able to answer “reversed” questions that could not be solved by a classical algorithm.

Another group [10] described a Chinese calendar savant who had exceptional proficiency in converting a date from the Gregorian calendar to the Chinese calendar. The results did not support the hypotheses of rote memorization, eidetic imagery, high speed calculation and anchoring strategy, use of calendar irregularities or monthly configurations. The authors proposed that this particular savant was familiar with 14 calendar templates and the knowledge of

matching these templates to every year. For dates beyond the 20th century the calculation was accomplished by regressing the date to a corresponding year in the 20th century by adding or subtracting 28 or 700 years. This explanation from the authors may be difficult for the reader to follow and it would seem even very much more difficult to put the calculations into play to accomplish this conversion of dates.

Other explanations for the CC have not been so complex, but neither have they been very informative. Pring and Hermelin [11] concluded that savants adopt a cognitive style of weak central coherence, protecting single representations from being retained in the form of stable enduring wholes. This strategy allows for transformation, reorganization and reconstruction of the relationship between single items of information. In other words, savants are more likely to process details at the expense of global information. Casey et al. [12] discussed the dysfunctional attention hypothesis to explain the skills of CC by the ability to divide, shift, direct and sustain attention. They concluded that a deficit in shifting selective attention from one stimulus to another was attributed to an inability to disengage attention as a result of a deficient orientation and an overselectivity. In other words, this latter explanation may come down to a focused attention. Finally, Hermelin and O'Connor [13] concluded that these human calculators used rule-based strategies. One last "explanation" was given by another group [9], suggesting that the CC are "solving the problems in a non-algorithmic way", meaning that no logical set of rules are followed in arriving at any answers. It may be helpful to indicate how the savants do not perform their skills, as one step closer to understanding how they do perform these same skills.

3.1.2 Musical savants

Absolute or perfect pitch is the ability to properly identify any musical note after hearing it. On a test involved with absolute pitch, investigated by Pring et al. [14] the savants scored higher than non-musicians, but not statistically better than a group of professional musicians. Also, in short-term memory tests for musical phrases the savants and musicians performed indistinguishably. The authors concluded that the skills of musical savants are separate from general intelligence, but also confirming that absolute pitch can, in fact, exist in these individuals. According to Heaton et al. [15] the autistic savants performed in a highly superior way, compared to controls who had self-reported absolute pitch. Five years earlier, Heaton [16] included other data, claiming that the savant group showed enhanced pitch memory and pitch labeling and also superior chord segmentation. Young and Nettelbeck [17] reported on a savant with perfect pitch, exceptional recall and performance of structured music and also an increased speed of information processing. Although this savant had high levels of concentration and memory, he had difficulties in verbal reasoning.

3.1.3 Hyperlexia

The life of one of the most famous savants, Kim Peek, was dramatized in the popular movie, "Rain Man", played by actor Dustin Hoffman. Kim reads the left side of a page with his left eye and simultaneously the right side of the page with his right eye (without a corpus callosum). The time taken for these two pages for Kim is usually 8 seconds and upon testing for retention he was 99% correct of the material just read [2]. These values are in contrast to 45 seconds for reading and 45% correct on testing seen in a group of normal individuals. Goldberg [18] described the condition of hyperlexia that the skill arises without any practice, is not integrated with other areas of knowledge, is associated with a dysfunctional

procedural memory system, but intact declarative memories. Finally, O'Connor and Hermelin [19] described two autistic children whose reading speeds were considerably faster than that seen in controls. To these investigators efficient grapheme-phoneme (written-sound) conversion is likely a modular component of the reading skill responsible for the hyperlexia in these savants.

3.1.4 Various other skills

One group [20] mentioned hypermnesia as one of the special skills found among those with the SS. An excellent example is Kim Peek who remembers nearly everything that he has read in 9,000 books [2]. That same group also mentioned several branches of the arts, as part of the SS, such as drawing, painting and sculpturing. Mottron and Belleville [21] described one individual who could draw objects rotated in 3-dimensional space more accurately than controls. In addition, this individual could detect a perspective incongruity between an object and a landscape at a superior level. The authors claimed that this skill was reached without the use of explicit or implicit perspective rules. Two years earlier the same authors [22] described the same patient with the same outstanding ability for 3-dimensional drawing. Their complex conclusion (difficult to understand) was that the performance was an “anomaly with hierarchical organization of the local and global parts of a figure, a local interference effect in incongruent hierarchical visual stimuli, a deficit in relating local parts to global form information in impossible figures and an absence of feature grouping in graphical recall”.

O'Connor and Hermelin [23] discussed gifted artists who were outstanding in the perception of a recognition memory for shapes. Results indicated that reproduction ability depended on artistic ability, independent of intelligence. Somewhat related to the skill of drawing was the talent of a 4-year-old boy [24]

who had an exceptional memory for maps and spatial locations, although the boy had a low-average intelligence and impaired motor skills. Luszki [25] mentioned a savant who was excellent on a block design test, but scored only 76 on a performance IQ test. Dowker et al. [26] described a savant poet who referred more often to aspects of self-analyses, while descriptions of other people not related to the self were less frequently mentioned. This latter emphasis on self is consistent with one of the basic core features of autism. Another special talent is the mechanical ability to repair everything and anything. Brink [27] described such a mechanical genius with a right-sided paralysis and transient loss of speech from a bullet into the left temporal area. Hoffman and Reeves [28] reported on a similar savant who could not speak until 20 years of age, but had great mechanical ability. Finally, other investigators [29] have described artists who were savants, commenting that their specific ability can occur at any level of intelligence, usually between 30 -75 IQ levels.

From all of these examples, it seems clear that nearly any specific skill or talent can occur in the SS.

3.2 Acquired SS

3.2.1 Without seizures

Treffert [30] has written extensively about the congenital form of SS, likely present at around birth, but also he has discussed the acquired form occurring later in life. In this instance “after some brain injury or brain disease, savant skills unexpectedly emerge, sometimes at a prodigious level, when no such skills were present before injury or illness”. Many examples were given by Treffert, including a 10-year-old boy knocked unconscious by a baseball, then later could do quick calendar calculations, an 8-year-old boy with the similar talent after a left hemispherectomy and a 3-year-old child after meningitis was later considered a

musical genius. Also included was a 9-year-old boy who was shot with a bullet to the left brain, leaving him with a right-sided hemiparesis, later developing special mechanical abilities. Finally, two painters were mentioned who had significant qualitative improvements after strokes involving the left occipital lobe and thalamus.

A surprising number of acquired cases had been earlier diagnosed with fronto-temporal dementia (FTD). These cases included a 68-year-old man without any interest in art who became an accomplished artist after FTD and also two other similar patients. In addition, another case was an example of “paradoxical functional facilitation” as a release phenomenon “where loss of some skills permits emergence of others”, like artistry. One investigator, mentioned by Treffert [30], had collected 12 cases of FTD with later emergent artistic ability. One particular patient was a 51-year-old artist whose skills surfaced for the first time after a subarachnoid hemorrhage associated with cerebral artery aneurysms. The frontal damage in this latter patient was similar to that seen in FTD. Finally, a talented art teacher at age 49 years at the beginning of a progressive FTD process, later showed an “impressive artistic growth”.

3.2.2 With seizures

Only very few, if any, examples can be found in the world literature of acquired SS with epilepsy. One possible example comes from Oliver Sachs [31] who described a person with “high fever, weight loss, delirium and perhaps seizures”. Following this illness this individual began painting extremely accurate scenes that colleagues believed could not have been painted before the illness.

The case of Patrick Obyedkov, a 35-year-old musical savant after the onset of seizures, was a fictional character on the TV serial, “House”.

The example of Daniel Tammet (D.T.) should be mentioned, but may not be relevant here, because his childhood seizures at 3 years of age had always been considered as not related to the SS but instead to his synesthesia. The latter condition occurred after his seizures. For only 3 years Daniel was on anti-epileptic drugs and he has been seizure free since early childhood [32]. He experienced this synesthesia in the form that every number that he experiences has its own color, shape and texture. At 4 years of age he developed rapid calendar calculating ability, in addition to an incredible memory for numbers. On American TV many viewers witnessed Daniel at 26 years of age in front of Oxford University dons reciting (without a single mistake) the value of Pi to 22,514 decimal places over a 5-hour period. Thus, this prodigious memory was not likely related to his early seizures at 3 years of age. Since nearly one-half of savants have A, which is closely associated with seizures, speculation would be the savants might have seizures, but they usually do not have such attacks.

Thus, only one case of possible seizures can be found in the world literature of either the acquired SS or the congenital type. On the other hand, many examples can be found of acquired SS without seizures, but with some damage or disturbance to the brain, usually the left frontal temporal area. Since a seizure condition involves a hyperexcitable brain, this condition is not likely the kind of circumstance to release savant skills and this reviewer is not aware of any patients with a left temporal lobectomy who have later developed the SS. However, a hypoexcitable left frontal-temporal area from a head injury, CVA, FTD, etc. seems sufficient to release the normal right hemisphere to develop the SS, escaping the “tyranny of the left hemisphere”.

One way to escape this “tyranny” is to stimulate the left frontal-temporal area with repetitive magnetic transcranial pulses. Snyder et al. [33] reported that 8

of 12 normal individuals improved their ability to accurately guess the number of discrete items that were placed together. In a study by Young et al. [34] 5 of 17 subjects developed savant types of skills, including declarative memory, drawing, mathematical and calendar calculating during the stimulation. In another study published 3 years earlier Snyder et al. [35] reported that 4 of 11 subjects displayed enhanced proofreading ability during the magnetic pulses. These findings argue that all of us under certain circumstances, like inhibiting the left frontal-temporal area may have the possibility of savant types of skills! This possibility should stimulate the readers to consider the potentiality of their own cognitive powers or artistic talents.

4. Possible causes of the SS

Takahata and Kato [36] have discussed the acquired SS, exemplified by patients with FTD who then develop savant skills, and have provided a few explanations to account for this phenomenon. First is the hypermnesic model that these skills develop from existing or dormant cognitive functions, like memory. Second is the paradoxical functional facilitation model, which emphasizes the role of reciprocal inhibitory interactions among cortical regions. Third is the autistic model, including the weak central coherence theory, also emphasizing the underconnectivity or disruption of long-range fibers, but with an enhanced local connectivity in given areas. The authors speculated that the enhanced local connectivity results from the specialized and facilitated cognitive processes responsible for savant skills.

The hypermnesic model with skills developing from existing functions finds some support from Miller [37] who concluded that skills exhibited by savants shared many characteristics found in normal subjects and that the skills are usually accompanied by normative levels of performance in various subtests. Also, this investigator claimed that it is unclear that savants have distinctive cognitive strengths or motivational dispositions. Pring and Hermelin [38] similarly

concluded that there may be no differences between savants and normal subjects regarding the nature of mental structures underlying specific talents, which are independent of the level of general cognitive functioning. These latter 2 studies do not therefore clarify how the savants perform their outstanding skills.

The third autistic model is based, in part, on the weak central coherence theory, which some authors [39] believe would predispose individuals to develop their talents with obsessive preoccupation. Others [40] have drawn a similar conclusion, that the excessive use of cognitive processing would be “instigated by the probable failure of central executive control mechanisms”. Still others [11] concluded that a “cognitive style of weak central coherence may protect single representations from being retained in the form of stable enduring wholes”. Happe [41] also concluded that a cognitive style biased towards local rather than global information processing (called weak central coherence) may be involved in the SS. Also, other authors [22] have referred to an anomaly in the hierarchical organization of the local and global parts of the skill involved. The other part of the autistic model is the presence of long-range fiber underconnectivity, emphasized by Hughes [42], also associated with enhanced short-range or local connectivity.

Many other explanations have been suggested with various depths of insight into the fascinating phenomenon of the savant syndrome. One group [43] hypothesized that a mutation gives rise to the development of the positive aspects of the SS, but has a deleterious effect on the development of other phenotypic traits, resulting in autism. Nurmi et al. [44] reported that chromosome 15q11-q13 involving the GABRB3 gene provided a genetic contribution to a subset of affected individuals with savant skills. Two years later Ma et al. [45] failed to demonstrate a linkage to 15q11-q13. Casanova et al. [46] had an interesting approach to the possible explanation of these skills by an analysis of the morphometry of 3 distinguished scientists who died and had an autopsy. They reported significant differences in these scientists' cortex from

comparison groups in both smaller minicolumn width and mean cell spacing. These conditions would be expected to enhance neuronal processing, as would be anticipated in the SS.

Mottron et al. [47] presented the concept of “enhanced perceptual functioning” to explain the SS. Included were possibilities of locally oriented visual and auditory perception, enhanced low-level discrimination and the use of a more posterior network in complex visual tasks. Also included were enhanced perception of static stimuli, decreased perception of complex movement, autonomy of low-level information processing toward higher-order operations and increased perceptual expertise. Consistent with the complexity of the latter explanations, Kelly et al. [48] found no evidence that the savants used short-cut strategies.

The “dysfunctional attention hypothesis” of Casey et al. [12] addressed the deficiencies of savants in shifting selective attention from one stimulus to another. This hypothesis does not clarify the mechanisms used by savants to explain their talents, except to imply that savants are locked into one stimulus. The 3 hypotheses offered by Kehrer [49] were that savants have a different perception and storing of perceived impulses, an abnormal memory process and storing function and these outstanding abilities are reinforced by the environment. However, these points do not easily explain the positive aspects of the SS. O’Connor and Hermelin [50] claimed that savants use strategies founded on “deduction and application of rules governing the material upon which their special ability operates”, also generating “novel examples of such rule-based structures”. Perhaps, these comments are not too helpful in understanding the SS, but another point is more helpful that these savants have an obsessional pre-occupation with a limited section of the environment. The theory of Goldberg [18] was that savants have a dysfunctional procedural memory system (related to skills or procedures) though their declarative memories (of facts) are intact. O’Connor and Hermelin one year earlier [51] than the later report [50] concluded that the savants have a superior image memory and ready access to a ‘picture lexicon’. The example of savants like Daniel Tammet, who can quickly multiply a long number by itself, requires comment. It is as if the problem is presented to these savants and they ‘read off’ the

answer as the brain ‘automatically’ does the calculation. While listening to a familiar opera, this reviewer often ‘automatically’ knew the next note, but this knowledge derives from learning the sequence of notes after many times hearing those same sequences. The parallel with the SS is that they also deal with sequences, but the major difference here is that the savants show these prodigious skills without any rehearsal of the problem just presented to them.

5. Characteristics of savants

5.1 Intelligence

The intellectual functioning was viewed as “average” [52], or “limited” [5, 48], “independent” [50, 51, 53, 54] and also “low average” [24]. O’Connor [55] has added that all savants can abstract to some degree.

5.2 Memory

Memory for the specific talent was described as exceptional [24], but as normal for general and verbal memory, independent of the measured verbal IQ [56]. Also, skills were not based on efficient rote memory [50]. Another characteristic is that skills of savants do not change with practice [50, 55].

5.3 Sensation and perception

As previously mentioned, Mottron et al. [47] proposed “enhanced perceptual functioning” in savants. Included are the possibilities of locally oriented perceptions, use of a more posterior network, enhanced perception of static stimuli, decreased perception of movements and differences between perception and general intelligence. These conditions add up to increased perceptual processing . On the other hand, Chen et al. [57] reported normal auditory and visual evoked potentials in savants.

5.4 Focus

Bor et al. [58] described one savant under discussion as having a “propensity to focus on local detail”. Casanova et al. [46] had a similar phrase, a “focused attention”,

likely related to an “obsessive personality profile” [40]. Hou et al. [59] stated this characteristic as a “focus on one topic at the expense of other interests”, similar to the “preoccupations” of O’Connor and Hermelin [50, 51, 53].

6. Anatomical relationships

One of the most fascinating and significant points in this review has been emphasized by Treffert [29] who has summarized the condition of an acquired SS. He has stated that damage to the left frontal-temporal area seems required to escape the “tyranny of the left hemisphere” and thus to permit the development of a savant skill by the right hemisphere. After studying all types of the SS, Bujas-Petkovic [60] concluded that “the functioning of the right cerebral hemisphere is most probably responsible for those abilities”. Treffert [1] has summarized the situation in another way, by concluding that left-brain damage occurs with right brain compensation. As previously mentioned, Snyder et al. [33] was one of the investigators who reported inhibiting the left anterior temporal area by repetitive magnetic stimulation, could result in the improvement of various abilities. Hou et al. [59] agreed that “the anatomic substrate for the savant syndrome may involve the loss of function also in the left temporal lobe”, but added that there is also enhanced function of the posterior neocortex.

The prefrontal area has also been emphasized in some reports. Takahata and Kato [36] referred to the paradoxical functional facilitation model in which reciprocal inhibitory interactions occur among adjacent or distant cortical regions, especially that of the prefrontal cortex and the posterior regions of the brain. The same prefrontal area was mentioned by Bor et al. [58] reporting on a celebrated savant (D.T.) whose fMRI showed hyperactivity in the lateral prefrontal cortex when demonstrating one of his skills by encoding digits.

7. Incidence

Treffert [1] has summarized data on the incidence of the SS. As previously mentioned, he has stated that approximately 10% of autistic persons exhibit savant abilities, that roughly 50% of those with the SS have autism and the remaining 50% have other forms of a developmental disability. The remaining cases of the SS include the acquired group that was previously discussed. The same author indicated that only 50 – 100 individuals in the world have these exceptionally prodigious talents. One other report [61] judged the general incidence of the SS in Finland, concluding that there were 45 cases in that country with an incidence rate of 1.4/1000 (1 in 714) who also had mental retardation.

8. Summary

The goal of this report is to review the Savant Syndrome (SS) and to explore its possible relationship to epilepsy, especially because nearly one-half of savants are autistic and many autistic individuals have seizures. There are 2 different forms, congenital and acquired. Among the congenital types, calendar calculators are one of the most common forms and one individual could provide the day of the week for any past event in 700 msec!. The explanations for the way in which this is done have varied from the simple to the complex. Some claim that a 'weak central coherence' is associated with a mental bias toward details at the expense of global information and others have suggested that the process involves complex matching templates to every year also with regression of dates. Musical savants with absolute or perfect pitch have been identified, at times with superior chord segmentation and increased speed of information processing. Hyperlexia is another form, exemplified by the 'Rain Man', Kim Peek, who reads the left side of a page with his left eye and simultaneously the right side with the right eye in 8 seconds with a 99% correct recall. Various other skills include hypermnesia, 3-D drawing, map memory, outstanding poetry, painting and all types of other artistic talents.

The acquired form refers to the development of outstanding skills after some brain injury or disease. Examples include a number of individuals with meningitis, bullets in their brains, CVAs, etc. who later develop savant skills. Also, savants who have had fronto-temporal dementia, usually involving the left side are found and represent an example of the acquired type, releasing the right hemisphere to develop savant skills by avoiding the 'tyranny of the left hemisphere'. Only one possible case of seizures could be found in the world literature associated with the savant syndrome and a hyperexcitable cortex with a seizure disorder does not seem to be compatible with a SS. In order to escape this tyranny of the left fronto-temporal area, transcranial magnetic stimulation inhibiting this area has led to nearly one-half of normal subjects developing savant types of skills during the actual stimulation. This finding suggests the great potentiality that all of us may have in cognitive powers or artistic talents if conditions were right for these skills to emerge.

The stated causes or explanations of the SS have been varied, including a 'paradoxical functional facilitation', but also an enhanced local connectivity as a compensation to the underconnectivity of long-range fibers. This latter point is usually mentioned in conjunction with a weak central coherence with attention to detail rather than the stable whole. Other explanations include an 'enhanced perceptual functioning' or a dysfunctional attention or an obsessive pre-occupation.

Characteristics of savants include average or limited intelligence, exceptional memory of their special talent, but otherwise normal without any change in the talent with practice. Also included is a propensity to focus on local detail.

The anatomical relationships involve the development of skills in the right hemisphere after inhibiting the damaged left fronto-temporal area. Also, the prefrontal area may be special by the hyperactivity noted in that same area while performing the skills.

The incidence of the SS is that approximately 10% of autistic persons exhibit savant abilities. Roughly one-half of the SS are autistic and the rest have other forms of developmental disability. One new and exciting category is the acquired SS.

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