

For this reason, linguistic processes are often formalized as a pair, consisting of a structural description and a structural change. Modern constraint-based theories have replaced this kind of formalization by a more modular approach in which very general operations are kept in check by a collection of abstract principles.

A final word of caution. Generative linguists take linguistic structures to be mental objects and assume

that these structures are formed and modified by linguistic processes of some sort. However, because linguistic theories generally abstract away from actual language processing, it is an open question to what extent linguistic processes are a direct reflection of mental computation.

See also: Levels of Adequacy, Observational, Descriptive, Explanatory; Rules and Principles.

Sturge–Weber–Dimitri Syndrome and Language

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The association of cerebral leptomeningeal angioma and facial nevus flammeus in the territory of the first branch of the trigeminal nerve ipsilateral to the angioma is known as Sturge–Weber syndrome (SWS). The cases with absence of a facial angioma are usually considered to be variants of the syndrome. This is a rare congenital neurocutaneous syndrome of unknown origin that occurs in both sexes with a frequency of approximately 1 per 50 000 (e.g., Comi, 2003). Sturge first described this disorder in 1879. In 1922, Weber described a similar clinical case and advocated the use of the term ‘encephalotrigeminal angiomatosis’ to describe it. It is characterized by leptomeningeal angiomatosis, glaucoma, and ipsilateral facial capillary hemangioma (port-wine stain) in the ophthalmic division of the trigeminal nerve (Aicardi, 1992). The facial nevus flammeus is present in 98% of children with SWS and it is visible at birth (Sujansky and Conradi, 1995b). It usually affects the upper eyelid and the forehead. The stain is caused by an overabundance of capillaries just beneath the surface of the affected skin. The angioma is typically located on the parietal and occipital lobes. It has been suggested that the angioma results from the failure of the primitive cephalic venous plexus to regress and properly mature in the first trimester of development (Comi, 2003). Normally, this vascular plexus forms in the sixth week and regresses at approximately the ninth week of gestation.

SWS is also characterized by neurological manifestations, which include focal or diffuse seizures (Kramer *et al.*, 2000), abnormal cerebral glucose metabolism (Lee *et al.*, 2001; Maria *et al.*, 1998; Pfund *et al.*, 2003; Pinton *et al.*, 1997), enlargement of the choroid plexus (Guermazi *et al.*, 2000), progressive atrophy and calcification of the brain (Comi, 2003;

Jay, 2000; Shamoto and Chugani, 1997), and recurrent stroke-like episodes (see Thomas-Sohl *et al.*, 2003, for a review of clinical symptoms and management). Hemianopia, progressive hemiparesis, motor deficits, developmental delay, and mental retardation are other dysfunctions associated with this neurological disorder. Mental retardation is present in approximately 50% of children with SWS (Sujansky and Conradi, 1995a) and clinical severity of signs and symptoms varies widely from one patient to another (Maria *et al.*, 1998).

It has been hypothesized that two main complications of SWS may explain the progressive neurological deficits associated with the disorder: the occurrence of seizures and cerebral ischemia. Thus, repeated seizure activity is associated with developmental delay, permanent hemiparesis, and mental retardation. In addition, venous stasis and recurrent episodes of venular thrombosis may be responsible for neurological deteriorations (cerebral calcification, gliosis of the cortex and white matter) and postnatal hemispheric atrophy (Comi, 2003; Kramer *et al.*, 2000; Portilla *et al.*, 2002).

Epilepsy affects over 80% of the children with SWS (Sujansky and Conradi, 1995a; Ville *et al.*, 2002). In a majority of children, onset occurs during the first year of life. Of the children who did not develop seizures during their first 2 years of life, only 14% developed seizures subsequently. It has been suggested that onset of seizures during the first 2 years of life and the presence of uncontrollable seizures are responsible for the developmental deterioration of previously normally functioning children with SWS (Roach, 1992). Thus, according to Sujansky and Conradi (1995a), later age of seizure onset is a favorable prognostic indicator. Contrary to these observations, Kramer *et al.* (2000) showed that cognitive delay is significantly correlated with seizure intensity, rather than the age of onset.

When seizures are unihemispheric and intractable to medication, hemispherectomy is strongly

recommended. This type of surgery is used in an attempt to control seizures and to prevent neurological deterioration (Kossoff *et al.*, 2002). Generally, bilateral angiomas is a contraindication to epilepsy surgery because a more diffuse region of epileptogenesis, which is not resectable, is more probable and the seizure prognosis may thus be unfavorable (Arzimanoglou *et al.*, 2000; Roach *et al.*, 1994). Nevertheless, according to some authors, epilepsy surgery in SWS should not be used too restrictively (Tuxhorn and Pannek, 2002). Tuxhorn and Pannek presented two bilateral cases of SWS that showed good seizure control and remission of epileptic encephalopathy after functional hemispherectomy.

Language development in SWS has not been well studied. Nevertheless, a few case studies have shown that language impairments associated with this syndrome result principally from epilepsy and following hemispherectomy. Vargha-Khadem *et al.* (1997) described the case of a 9 year old boy diagnosed with SWS affecting the left hemisphere, in which seizure activity is the cause for arrested speech development. This boy, named Alex, experienced his first epileptic episode 6 days after birth. Despite having normal hearing, Alex did not develop speech, and his utterances, at 33 months of age, were still limited to babbling. Furthermore, his intellectual development was globally delayed. Between the ages of 4 and 8 years, Alex attempted to communicate with gestures. At age of 8 years 6 months, Alex underwent a successful left hemispherectomy. Interestingly, 10 months after surgery, Alex suddenly began to develop speech and, by age 10, could converse with copious and appropriate speech. This child achieved remarkable linguistic competence with an isolated right hemisphere and he speaks clearly and fluently with grammatically correct sentences. However, in the domain of receptive language, Alex showed poor comprehension of more complex grammatical sentences, as assessed by the Token Test (De Renzi and Vignolo, 1962) and the Clinical Evaluation of Language Functions-Revised (CELF-R; Semel *et al.*, 1987). Other cognitive dysfunctions have been noted in Alex's case, such as reading, writing, and arithmetic. At age 13, his performance in these cognitive areas was still at the basal level (i.e., <6 years).

Mariotti *et al.* (1998) described the linguistic abilities of a woman with SWS who underwent early removal of her left hemisphere, at age 3. Extensive assessment of her linguistic abilities was conducted when she was 20. In contrast to Alex's case, her performance, when compared to mentally retarded controls matched for age, education, and IQ, showed fluent spontaneous speech with intact pragmatic abilities, normal articulation, prosody, and phrase length, and grammatically correct sentences. Reading and

writing on copy were also normal, although writing on dictation was slightly impaired. As in Alex's case, however, she showed a marked impairment in more complex syntactic comprehension of sentences, low IQ, and poor memory span.

These observations in the case of SWS and hemispherectomy indirectly suggest a necessary integrity of the left cerebral hemisphere to adequately process morphosyntactic aspects of language. However, not all authors agree with this interpretation on the issue of cerebral hemispheric specialization of function (e.g., Bishop, 1988; Mariotti *et al.*, 1998; Vargha-Khadem *et al.*, 1997). Rather, they attribute the observed deficits to a low IQ and limited verbal memory span in children with SWS. Mariotti *et al.* (1998) suggested that failure of syntactic tasks could be merely the expression of a generic cognitive impairment (such as poor lexical knowledge) and not of specific linguistic damage. Thus, it is unclear whether the language dysfunction seen in SWS can be ascribed to a low IQ and poor working memory.

See also: Anatomical Asymmetries versus Variability of Language Areas of the Brain; Language in the Nondominant Hemisphere; Language Development: Overview.

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Sturtevant, Edgar Howard (1875–1952)

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Edgar H. Sturtevant was born in Jacksonville, Illinois. He studied at the University of Chicago (receiving his Ph.D. in 1901 with a dissertation on Latin case forms) and later became an assistant professor of classical philology at Columbia University in New York. In 1923 he joined the linguistics faculty at Yale University, which soon acquired a reputation in the United States for its strong linguistics program. In 1924, he was a member of the organizing committee for the founding of the Linguistic Society of America (LSA), in which Leonard Bloomfield and George M. Bolling actively participated.

Sturtevant played a major role at the Linguistic Institutes in the promotion of the study of phonetics, research on Native American languages, and field work on the Modern American English dialects.

Most of Sturtevant's works focus on historical linguistics and classical philology. When it was speculated that the Hittite language (ca. 2000 to 1200 B.C.) was related to the Indo-European language family,

Sturtevant became especially absorbed in the investigation of its genealogy. In this endeavor, as was customary at the onset of modern scientific linguistics of that time, the recovery of prehistoric linguistic systems and regularity of linguistic change were sought by the comparative method. His first major paper on this subject was 'On the position of Hittite among the Indo-European languages,' published in *Language* (1926). His position was that Hittite exhibits more archaic traits than the normally reconstructed forms for Indo-European, and hence both Hittite and Indo-European have an earlier common ancestor stage, namely, Indo-Hittite. On the other hand, it was shown that Hittite could be grouped into the Indo-European family after it was demonstrated by Kuryłowicz in 1927 that the language preserved distinct reflexes of the segments postulated in de Saussure's internal reconstructive hypothesis. Sturtevant dedicated himself to the phonological and morphological investigations of the Hittite language.

It should be stressed that while most linguists in the United States at that time were students of historical philology, Sturtevant was a significant figure who contributed to the shift from historical to non-historical