RECIPROCAL NEUROLOGICAL DEVELOPMENTS OF TWINS DISCORDANT FOR HYDROCEPHALUS

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In 1876, Galton reported studies of 35 sets of twins of 'close similarity' and 20 of 'marked dissimilarity' as the basis for defining and differentiating genetic *vs.* environmental factors determining human anatomy, physiology, intelligence and behaviour. He concluded that '... nature prevails enormously over nurture when differences in nurture do not exceed what is commonly found among persons of the same rank of society and in the same country'.

Subsequent studies of twin development have supported Galton's emphasis of the importance of genetic factors. For example, remarkable twin concordances have been reported for intellectual functions. A review of 52 studies (Erlenmeyer-Kimling and Jarvik 1963) revealed increasing inter-pair correlations of intelligence with increasing genetic similarity. Median correlations were low (0·23) for unrelated children reared together, higher (0·50) for parent-child comparisons, higher still for dizygotic twins (0·52), and highest for monozygotic twins reared apart (0·75) and together (0·87). A more recent review of the literature (Bouchard and McGue 1981) reported similar findings of systematic increasing correlations of intelligence with increasing genetic similarity.

In addition to development of intelligence, studies of monozygotic twins have also shown high concordance for a wide range of abnormalities, including schizophrenia (Gottesman and Shields 1972, Belmaker *et al.* 1974), phobias (Eckert *et al.* 1981), learning disorders (Hallgren 1950, Bakwin 1973), antisocial behavior (Christiansen 1970, Centerwall and Robinette 1989), depression (Allen 1976), stuttering and tics (Godai *et al.* 1976), and even of neuropathological processes developed in later life (Fairburn 1973).

Studies of hydrocephalus in twins have reported levels of concordance ranging from 7·8 per cent (five of 59 like-sex twins), based on 15 studies world-wide, to 21 per cent (16 of 76 like-sex pairs), based on Japanese birth records between 1969 and 1985 (Imaizumi 1989). Interpretation of these studies is qualified by the lack of definitive zygosity determinations, failure to document the type of hydrocephalus and, perhaps most significantly, possible contributions of non-genetic factors, such as twin-related birth complications, which may lead to development of hydrocephalus in one or both twins.

Other studies focusing on differences between twins have supported Galton's disclaimer, *i.e.* that nature may not prevail over nurture when differences in nurture exceed unspecified environmental influences that he described as the norm. For example, monozygotic twins sharing

a common chorion have significantly higher inter-pair IQ correlations than monozygotic twins developing in separate chorionic sacs (Melnick *et al.* 1978, Rose *et al.* 1981). Moreover, numerous studies have documented evidence that pre-, peri- or early postnatal complications, including low birthweight, toxemia, preterm birth, umbilical cord tangling, and fetal vascularization and other congenital anomalies occur more frequently in twins than singletons (Benirschke 1972, Leetz 1976, Leroy 1976). In the light of the evidence of increased risks in twin births, reports of a higher incidence of learning disabilities and left-handedness in twin than in singleton births are not surprising (Record *et al.* 1970; Mittler 1971, 1976; Bakwin 1973). Other studies have reported systematic anatomical and functional differences in some monozygotic twins, which have been described as 'mirror-imaging' phenomena (Carter-Saltzman 1979, Gedda 1981). These twins are thought to be products of late embryonic splitting; not anatomical duplicates, but mirror-images of each other. Such twins show mirror-imaging of facial asymmetries and reversed hair-whorl direction. Reported functional mirror-image phenomena include reversed hand-preferences and reversed ear-preferences for dichotically presented verbal material.

These accumulating findings indicate the importance of previously unsuspected intra- and extra-uterine environmental influences that apparently can variously modify the expression of genetic endowment. They are also consistent with the conclusions based on a review of the literature on heritability of intelligence: Bouchard and McGue (1981) noted the significance of non-genetic factors in the development of intelligence, as reflected by a 'large amount of unexplained variability within degrees of (genetic) relationship'.

The present study focuses on the significance of early environmental factors contributing to divergent outcomes for 10 sets of twins discordant for hydrocephalus.

Method
Comprehensive neuropsychological exam-

inations of 295 British children and young adults with hydrocephalus of early onset (Berker 1985) revealed 12 children who had non-hydrocephalic twins. Examination of 10 non-hydrocephalic twins provided opportunities for investigating similarities, differences and associated factors determining divergent outcomes in twins discordant for hydrocephalus. Although we identified 12 patients (4·1 per cent of the sample) who were products of twin births, in one set the co-twin had died, and in another permission to test the non-hydrocephalic twin was withheld. The remaining 10 sets of twins had complete examinations and their medical records were available.

Determinations of genetic identity were carried out in Britain by Cellmark Diagnostics of Abingdon. Analysis of blood samples taken from the eight like-sex twin pairs using the DNA finger-printing technique of Jeffreys *et al.* (1985) revealed six monozygotic and two dizygotic pairs. Thus our sample consisted of six monozygotic pairs, two like-sex dizygotic pairs and two opposite-sex dizygotic pairs.

Table I presents descriptive data for the 10 sets of twins. All 10 non-hydrocephalic children and eight of the 10 hydrocephalic children were attending normal schools. Seven were diagnosed as having 'congenital' hydrocephalus (*e.g.* no evidence of early pre- or perinatal intracranial hemorrhage, meningitis or other brain insults). Of the three remaining, two developed hydrocephalus following the onset of meningitis and one developed ventricular expansion following treatment of spina bifida (sacral myelomeningocele) shortly after birth. CT scans of the hydrocephalic children were obtained within two years before examination with the Michigan Neuropsychological Battery (MNB). The rationale and nature of the MNB are described in detail elsewhere (Campbell *et al.* 1981, Smith 1983).

CT scans of the hydrocephalic twins were classified according to criteria described previously (Lewin 1980, Lorber 1983). Three had gross hydrocephalus (CSF occupying over 70 per cent of the cranial cavity) and five had moderate hydrocephalus (CSF occupying 50 to 70 per cent of the cranial cavity). Two

TABLE I

Test performances and associated factors of 10 hydrocephalic (H) and their non-hydrocephalic (NH) twins

	1 VR H	JR NH	2 IB H	SB NH	3 IW H	NW NH	4 MB H	MIB NH	5 CY H	DY NH	6 HL H	JL NH	7 NM H	AM NH	8 RA H	AA NH	9 JH H	CH NH	10 SW H	CW NH
Zygosity[1]	DZ		DZ		DZ		DZ		MZ		MZ		MZ		MZ		MZ		MZ	
Age (yrs)	9		17		16		15		9		7		12		17		5		8	
Sex	F	F	M	F	M	M	M	F	M	M	F	F	M	M	M	M	M	M	F	F
SES[2]	3B		3B		2		1		3B		2		2		3A		3B		2	
School[3]	N	N	N	N	S	N	N	N	S	N	N	N	N	N	N	N	N	N	N	N
Scan[4]	Mod		Mod		Gross		Mod		Slit		Gross		Mod		Gross		Slit		Mod	
Shunt	+		+		+		+		+		+		+		+		+		-	
Revision	2		4		6		2		2		0		0		1		3		-	
Seizures	+		-		-		-		+		-		-		-		-		+	
Type[5]	Con		Men		Men		Con		Con		Con		Con		Con		Con		SM	
Hand[6]	L	R	L	R	R	R	R	R	L	R	L	R	L	L	L	L	L	R	L	L
Eye	R	R	L	R	L	R	L	R	L	R	L	R	L	R	L	R	L	L	R	R
Birthweight (g)	2168	2126	1814	1361	2325	2296	1956	2070	<907	907	2353	1758	2041	1985	992	1503	2495	2041	2041	2835
Gestation	Term		-4wks		Term		-5wks		-7wks		Term		-7wks		-2mths		-5wks		-2wks	
Order[7]	H 5		H 30		H 10		NH 5		H<1		H 5		H 5		H 5		H 10		H 30	
Mat. age (yrs)	19		30		32		26		28		29		26		23		31		26	
VIQ	62	88	77	92	82	99	113	109	65	79	77	96	81	111	96	100	102	124	96	118
PIQ	45	104	70	94	68	126	109	136	58	96	80	102	100	129	77	98	81	100	90	126
FIQ	50	94	72	92	74	112	112	125	58	86	76	99	89	122	87	99	91	114	92	124
SDW[8]	2	10	1	8	1	12	10	13	1	10	6	8	4	12	6	8	9	10	8	13
SDO[8]	1	11	2	8	3	13	10	18	1	11	3	12	6	13	6	8	7	8	10	13
Sensory[9]	B	N	1	N	1	N	N	N	B	N	N	N	N	N	N	N	B	B	B	N
Motor[9]	B	N	BR	N	R	N	BR	N	L	N	R	N	BL	N	B	N	L	N	B	N

[1]DZ—dizygotic; MZ—monoxygotic.
[2]According to classification of British Registrar General.
[3]N—normal; S—special.
[4]Dilatation: gross or moderate (mod); slit—shunt overdrainage.
[5]Con—congenital hydrocephalus; men—post-meningitic; SM—associated with a sacral myelomeningocele.
[6]Determined by writing.
[7]First-born by time (mins).
[8]Symbol Digit Modalities Test—written (SDW) and oral (SDO): substitution scores based on mean of 10 (SD 3).
[9]Single and Double Simultaneous Stimulation Test: N—normal; I—intermittent, or B—bilateral sensory deficits.
[10]Purdue Pegboard Test: N—normal; B—bilateral and/or R/L motor deficits.

patients had slit (collapsed) ventricles as a result of overdrainage after shunt treatment. Three had a history of seizures. The hydrocephalic twin was the first-born in nine of the 10 pairs.

Results

Consistent with earlier reports (Berker 1985), the intellectual development of the 10 hydrocephalic children ranged from markedly subnormal (e.g. Wechsler Full-scale IQ of 50) to above average (Full-scale IQ of 112). Only five had Verbal and/or Performance IQs in the average range. In marked contrast, all 10 non-hydrocephalic children had Verbal and/or Performance IQs in at least the average range, with five above average.

Previous analysis of the entire sample of 295 British hydrocephalic patients (Berker 1985) revealed more limited development in patients with lower socio-economic status (SES) and in those with a history of seizures than in hydrocephalic children from higher SES groups and without seizures. Consistent with this, of the 10 hydrocephalic twins, the two with most limited development (Full-scale IQs of 50 and 58) had seizure disorders and were from the lowest SES group. In contrast, in addition to being the only one of the 10 who was not first-born, the hydrocephalic child with the highest IQ (M.B., aged 15, with Full-scale IQ of 112) was in the highest SES category and had no history of seizures; correspondingly, M.B.'s twin sister had the best over-all outcome among the non-hydrocephalic children (Full-scale IQ of 125). It is also noteworthy that M.B.'s Full-scale IQ of 112 was achieved despite moderate hydrocephalus (CSF occupying 50 to 70 per cent of the cranial cavity). These findings are also consistent with previous reports documenting the development of superior adult intellectual capacities,
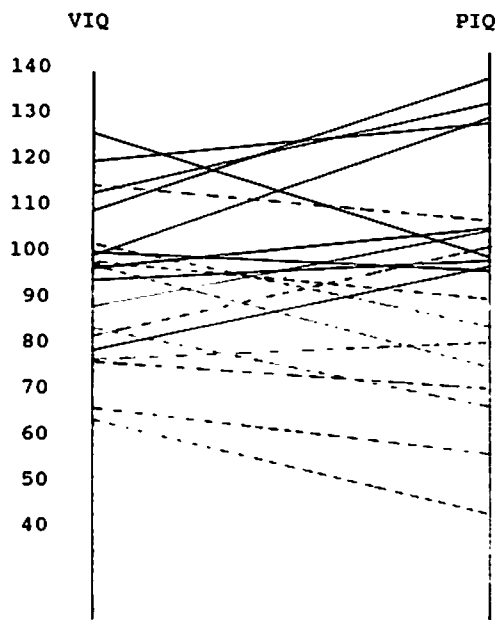
VIQ   PIQ

Fig. 1. Relationship between Verbal and Performance IQs for non-hydrocephalic twins (solid lines) *and hydrocephalic twins* (broken lines). *Predominantly positive slopes of solid lines indicate higher Performance than Verbal IQs for non-hydrocephalic twins* vs. *predominantly negative slopes and higher Verbal than Performance IQs for hydrocephalic twins.*

TABLE II
Significant differences between hydrocephalic (H) and non-hydrocephalic (NH) twins

	Group		p
	H	NH	
Mean VIQ	85·1	101·6	0·0256*
Mean PIQ	77·8	111·1	0·0005*
Left-handedness	8/10	3/10	0·0246**
Left-eyedness	8/10	2/10	0·0073**
VIQ>PIQ	8/10	2/10	0·0073**

*t test.
**χ^2 test.

hydrocephalic and non-hydrocephalic twins, using neuropsychological tests sensitive to organic cerebral dysfunction. Despite IQs in the average to above-average range for five of the hydrocephalic twins, all 10 had subnormal Purdue Pegboard performances (Costa *et al.* 1963), eight of whom also had subnormal Symbol Digit performances (Smith 1982), reflecting covert persisting effects of hydrocephalus, its etiological precursors and/or delayed emergent pathological sequelae. By contrast, all 10 non-hydrocephalic twins had normal performances on these two measures (Table I).

In addition to differences in degree of intellectual development, comparisons of the two groups revealed reciprocal patterns of development of verbal *vs.* non-verbal cognitive functions. Eight non-hydrocephalic twins had higher Performance than Verbal IQs (two to 27 points higher; mean difference 15·1 points). In contrast, eight of the hydrocephalic children had higher Verbal than Performance IQs (four to 21 points higher; mean difference 11·9 points). The pattern of greater development of verbal than non-verbal cognitive functions in the hydrocephalic twins *vs.* the reciprocal pattern in the non-hydrocephalic twins is illustrated in Figure 1. The reciprocal developmental patterns between the hydrocephalic and non-hydrocephalic twins are shown in Table II, which also shows that the difference between the mean Performance IQs (33·3) is more than twice as great as that between the mean Verbal IQs (16·5).

Hand and eye preferences provided additional evidence of reciprocal developmental patterns between the hydrocephalic and non-hydrocephalic pairs. Review of familial handedness showed right-hand preference in 19 of 20 parents. Consistent with common practice, all nine shunt-treated hydrocephalic children initially received right-hemisphere shunts, with subsequent revision to the left hemisphere in only one case. Despite an apparent genetic predisposition to dextrality and documented right-hemisphere neurosurgical interventions for shunt treatment, eight of the 10 hydrocephalic twins were left-handed, compared with

including cases with successful completion of university education, despite extreme hydrocephalus occupying up to 95 per cent of the cranial cavity (Dandy 1921, Lorber 1983, Berker 1985).

Previous studies of hydrocephalus and other early brain insults have documented limitations of IQ scores as reliable indices of development of over-all cerebral function (Prigatano *et al.* 1983, Smith 1983, Lezak 1988). Such limitations were demonstrated in comparisons of the 10

only three the non-hydrocephalic twins. The findings of systematically increased left-hand preference among hydrocephalic twins is similar to findings of increased left-eye preference among the hydrocephalic twins; eight of the hydrocephalic *vs.* two of the non-hydrocephalic twins (Table II).

In contrast to the 80 per cent rate of the left-hand preference for the 10 hydrocephalic children, the incidence of left-hand preference in our sample of 283 singletons with hydrocephalus was approximately 28 per cent (Berker 1985). This latter percentage is approximately the same as that for our sample of apparently normal non-hydrocephalic twins (30 per cent), and also of that reported in other studies of larger samples of apparently normal twins. The incidence of left-handedness in single-born hydrocephalic children and apparently normal twins is two to three times greater than that reported for normal singletons (Nachshon *et al.* 1983). Thus the extraordinarily high rate of left-handedness and eyedness for the hydrocephalic twins (eight of 10) apparently reflects pathological sinistrality due to cumulative effects of risk factors associated with twinning as well as hydrocephalus.

Of the eight left-handed hydrocephalic twins, two had right hemiparesis and a third had a congenital malformation of the right hand, leaving him with only half a right thumb and 1½ fingers. When the remaining five were compared with their non-hydrocephalic twins, two pairs showed evidence of mirror-imaging, in the form of facial asymmetry and reversed hair-whorl directions: left-handedness in these two hydrocephalic twins may reflect mirror-image phenomena. Thus for five of eight left-handed hydrocephalic twins, hand preference may have been influenced by early complications of hydrocephalus, developmental anomalies or mirror-imaging phenomena.

Discussion

Consistent with Galton's report over 100 years ago, this study demonstrates that, beginning with the formation of the zygote at fertilization and despite similar or identical genetic endowment, differences in nurture can contribute to strikingly divergent patterns of neurological development. In addition to discordance for hydrocephalus, the 10 sets of twins showed striking reciprocality in degree and nature of intellectual development, handedness, eye preference and birth order. These differences strongly suggest underlying systematic divergent developmental patterns in inter- and/or intrahemispheric cerebral organization.

Our finding that nine of the 10 hydrocephalic twins were first-born suggests that primacy in twins appears to be a risk factor related to development of hydrocephalus and associated left hand- and eye-preference. However, previous studies of hand preference and birth order, focusing on populations of apparently normal twins and singletons, have reported contradictory findings. Christian *et al.* (1979) studied 104 pairs of apparently normal twins discordant for hand preference. 84 per cent of the first-born monozygotic twins were left-handed, while dizygotic twins showed no effect of birth order on hand preference. A subsequent study of 82 pairs of twins discordant for handedness reported a greater incidence of left-handedness (by a factor of 1·8) in *second*-born twins (Boklage 1981). More recently, Searleman *et al.* (1989) reviewed the literature on the relationship between birth order, birth difficulties and lateral preference in apparently normal singletons. They reported 'no evidence to relate birth order position to deviations from right-sidedness'. However, they pointed out that 'more direct' measures of birth difficulties were significantly related to deviations from right-handedness.

Davis *et al.* (1987) reported a study of four sets of preterm twins discordant for post-hemorrhagic hydrocephalus, in which all four second-born twins developed hydrocephalus. These findings are exactly opposite to ours with respect to birth order and development of hydrocephalus. The apparently conflicting findings partly may reflect sampling differences. For example our study was not restricted to preterm infants, and none of our patients was diagnosed as

having post-hemorrhagic hydrocephalus. In addition, our twins constituted the first pregnancies and deliveries for each mother; Davis and colleagues did not specify the presence or absence of previous deliveries in their sample.

The rate of twinning in our sample of 295 hydrocephalic children (12 pairs or 4·1 per cent) is over three times the reported British rate of approximately 1·25 per cent (Mittler 1970). Since birth complications are more frequent among twins than singletons, the disproportionately high number of twins in our sample may reflect a greater number of twins than singletons who develop hydrocephalus following pre-, peri- or postnatal complications. Similarly, a significant proportion of twin pairs who are concordant for hydrocephalus may have both suffered birth complications rather than having a common genetic basis for their disorder. Thus attempts to establish the significance of heredity in the development of hydrocephalus based on twin concordance rates are qualified by high rates of birth complications among twins. Furthermore, the numerous increased risks reported for twin births and subsequent findings of impaired language and verbal development in this and other studies of apparently normal twins constitute important limitations, restricting the generalizability of twin research to normal intellectual development.

The systematic pattern of greater impairment of verbal than non-verbal functions in eight of the 10 non-hydrocephalic twins is consistent with previous reports by Mittler (1976) and others of an increased incidence of learning difficulties in *apparently* normal twins. Mittler reported only a 'weak' relationship between subnormal language development and birthweight, birth complications and length of gestation. Nevertheless, the increased pre-, peri- and early postnatal risk factors associated with twin births cannot be dismissed as coincidental or inconsequential.

The reciprocal pattern of impaired non-verbal reasoning capacities *vs.* relative sparing of language and verbal reasoning abilities in eight of the 10 hydrocephalic twins is consistent with previously reported studies of hydrocephalus (Dennis *et al.* 1981, Lonton 1984, Berker 1985), and with findings in patients with other types of diffuse bilateral cerebral involvement, including neurotoxicity, alcoholism, carbon monoxide poisoning, Alzheimer's disease and Cushing's disease, as well as with a normal decline in cerebrovascular and metabolic efficiency with advancing age (Whelan *et al.* 1980, Smith 1981, Berker and Smith 1988).

The developmental patterns of verbal *vs.* non-verbal functions and associated hand-preferences in both the hydrocephalic and normal twins are inconsistent with static doctrines of hemispheric specialization, which stress genetic pre-programming of the left hemisphere for development of language function and of the right hemisphere for visual-spatial function. According to the static model, the hydrocephalic group with greater relative development of language than non-language function would not be expected to have eight of 10 with left-hand preference, suggesting early compromise of the left hemisphere. Similarly, the non-hydrocephalic twins with greater relative impairment of verbal than non-verbal function would not be expected to have a near-normal incidence of right-handedness, suggesting patency of the left hemisphere. Instead, these findings suggest a more dynamic model of brain organization, disorganization and reorganization. Consistent with this, Davis *et al.* (1987) described a hydrocephalic twin with evidence of left-hemisphere pathology, who had visual-motor deficits in conjunction with apparently normal development of language function.

Longitudinal studies of children with hemispherectomy for intractable seizures have demonstrated that the neuroanatomical substrata necessary for the development of normal and even above-average or superior adult verbal and non-verbal intellectual functions are present in both hemispheres (Smith 1984). Studies of hemispherectomy and other types of cerebral insults have also revealed a functional hierarchy, in which reorganization of language and verbal functions usually takes precedence over visuoperceptual spatial reasoning capacities in patients with bilateral pathological

involvement of cerebral mechanisms (Smith 1972). This principle of cerebral reorganization is consistent with the outcome for eight of the hydrocephalic twins, as well as for other clinical groups with diverse types of diffuse brain insults.

However, this principle appears to be inconsistent with the outcomes of eight of the non-hydrocephalic twins, as well as of other apparently normal twins who have impaired language development and normal non-verbal reasoning capacities, without evidence of lateralized cerebral insult. Smith (1968) has suggested that patients with learning disability or impaired language development associated with normal non-verbal cognitive function usually have suffered bilateral damage. Thus persistence of impaired language development, despite normal non-verbal development, may reflect compromise or inhibition of specific cerebral mechanisms which normally mediate language function in the left hemisphere, as well as associated compromise or inhibition of compensatory mechanisms in the right hemisphere. It is noteworthy that none of the non-hydrocephalic twins showed evidence of residual lateralized left-hemisphere embarrassment on the Purdue Pegboard test of manual dexterity, the most sensitive test on the MNB to the presence of lateralized pathological involvement.

These findings suggest that although language mechanisms as a rule are resistant to the pathological effects of diffuse cerebral insults, in certain conditions (such as twinning) unidentified intra-uterine influences may selectively disrupt or inhibit bilaterally represented language mechanisms, while sparing bilaterally represented non-verbal visual-spatial mechanisms. Thus mechanisms present in each hemisphere that can mediate organization and/or reorganization of language and verbal reasoning abilities appear to be independent of—and may be structurally different from—bilaterally represented neural mechanisms that can mediate organization and/or reorganization of non-verbal functions.

Increasing refinements of positron emission tomography, magnetic resonance imaging and other radiological techniques in studies of intra-hemispheric cerebral organization of brain-behavior relationships may elucidate organizational differences in the nature and plasticity of language *vs.* non-language mechanisms within each hemisphere. Such studies may also contribute to our understanding of factors determining disorganization and reorganization of verbal *vs.* non-verbal functions following different types of insults incurred at various stages of cerebral maturation. Definitions of such factors may contribute to the heterogeneity and unexplained variability within degrees of genetic similarity noted by Bouchard and McGue (1981).

Obviously, the small size of our clinical population warrants no firm conclusions. Furthermore, we were not able to identify specific intra-uterine factors which selectively led to the development of hydrocephalus in only one twin, nor were we able to isolate pathological factors leading to the impaired development of language functions in the non-hydrocephalic twins. Nevertheless, this neuropsychological study of 10 twins discordant for early-onset hydrocephalus is strikingly consistent with other studies demonstrating the vulnerability of the infant brain to pre- or postnatal intra- or extracerebral insults, as well as its remarkable versatility and capacity for reorganization with cerebral maturation.

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SUMMARY

Studies of 10 sets of twins discordant for hydrocephalus in early life revealed striking differences in degree and nature of development of verbal *vs.* non-verbal cognitive functions, birth order, and hand and eye preference. Despite similar (four dizygotic pairs) or identical (six monozygotic pairs) genetic endowment and grossly similar intra- and extra-uterine environmental and socio-economic influences, the consistency of the differences between the hydrocephalic children and their seemingly normal twins indicate systematic differences in pre-, peri- and/or early postnatal organization and development of hemispheric function. Follow-up studies also documented development of above-average intelligence, despite drastically reduced cerebral mantle size in hydrocephalus of early onset. The atypical patterns of development of the non-hydrocephalic twins also confirm previously described qualifications reported in studies of the significance of genetic *vs.* environmental factors in twins.

RÉSUMÉ

Jumeaux discordants pour l'hydrocéphalie

Une étude portant sur 10 paires de jumeaux discordants pour une hydrocéphalie survenue en début de vie a révélé des différences frappantes dans le degré et la nature du développement des fonctions cognitives verbales *vs.* non-verbales, l'ordre de naissance et les préférences main-oeil. En dépit d'un programme génétique semblable (quatre paires dizygotes) ou identique (six paires monozygotes) et des influences intra- et extra-utérines, et socio-économiques globalement identiques, la consistence des différences entre les enfants hydrocéphales et leurs jumeaux apparemment normaux indique des différences systématiques dans l'organisation pré-, péri- et ou précocement post-natale et le développement de la fonction hémisphérique. Les études de suivi ont démontré la possibilité d'une intelligence au dessus de la moyenne, en dépit d'une réduction dramatique de l'épaisseur du manteau cérébral dans l'hydrocéphalie à début précoce. Les distributions atypiques du développement des aptitudes chez les jumeaux non hydrocéphales confirment également les points antérieurement reportés dans les études de signification des facteurs génétiques *vs.* environnementaux chez les jumeaux.

ZUSAMMENFASSUNG

Zwillinge bei denen einer einen Hydrozephalus hat

Untersuchungen von 10 Zwillingspaaren, bei denen einer einen Hydrozephalus hatte, haben auffallende Unterschiede bei der Entwicklung von verbalen *vs.* non-verbalen kognitiven Funktionen, Geburtenfolge und Hand- und Augen Präferenz ergeben. Trotz ähnlichem (vier dizygote Paare) oder identischem (sechs monozygote Paare) genetischem Material und weitgehend gleichen intra- und extrauterinen Umwelt- und sozioökonomischen Einflüssen, deutet die Übereinstimmung der Unterschiede zwischen Kindern mit Hydrozephalus und ihren offenbar normalen Zwillings-geschwistern auf systematische Unterschiede in der prä-, peri- und/oder frühen postnatalen Organisation und Entwicklung der Hemisphärenfunktion hin. Bei Kontrolluntersuchungen zeigte sich eine überdurchschnittliche Intelligenzentwicklung trotz deutlich verminderter Hirnrinde bei früh beginnendem Hydrozephalus. Atypische Entwicklungsmuster bei den nicht hydrozephalen Zwillingen bestätigen auch früher beschriebene Befunde, die in Untersuchungen zur Signifikanz von Genetik *vs.* Umweltfaktoren bei Zwillingen beschrieben werden.

RESUMEN

Gemelos hidrocefálicos discordantes

Estudios realizados en 10 grupos de gemelos discordantes en su hidrocefalia precoz revelaron diferencias notables en el grado y naturaleza del desarrollo de las funciones cognitivas verbales frente a las no verbales, el orden de nacimiento y la preferencia de mano y ojo. A pesar de una genética similar (cuatro pares dizigóticos) o idéntica (seis pares monozigóticos) y un entorno más o menos similar intra y extrauterino y similares influencias socio-economicas, la consistencia de las diferencias entre los niños hidrocefálicos y sus mellizos normales indica que hay diferencias sistemáticas en la organización pre-, peri- y/o postnatal de la función hemisférica. Estudios de seguimiento también documentan sobre un desarrollo de la inteligencia por encima del promedio, a posar de una reducción drástica del tamaño del manto cerebral en la hidrocefalia de inicio precoz. El patrón atípico del desarrollo de los gemelos no hidrocefálicos confirma también las calificaciones previamente descritas aportadas en estudios sobre el significado de la genética frente a los factores del ambiente en gemelos.

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