Diseases of the circulatory system encompass a wide spectrum of cardiovascular ailments including heart, circulation and cerebrovascular diseases. Heart and circulation ailments include rheumatic heart disease, hypertension, ischemic heart and pulmonary circulation diseases, pericarditis, functional heart disorders such as heart valve disfunctions, cardiomyopathy, and congestive heart failure. Cerebrovascular disorders include hemorrhages of blood vessels, arteriovenous malformations, arterial occlusions, aneurysms and other disorders affecting arteries, arterioles, capillaries, veins and lymphatics. Diseases of the blood and blood forming organs include anemias, inherited and acquired, coagulation defects and diseases of white blood cells. Congenital defects of the circulatory system include atrial and ventricular septal defects, coronary artery fistulae, and anomalies of the pulmonary artery.

There are 65 references in Chapter 13 (19.5% of the 334 references in Part Two)—48 annotated and 17 supplemental. Thirty-two references to remission of circulatory system diseases (ICD-9-CM* numbers 390-459), 13 references to remission of diseases of blood and blood forming organs (ICD-9-CM numbers 280-289), and 8 reports of resolution of congenital defects of the circulatory system (ICD-9-CM numbers 745-747) have been combined in this chapter. Full text of 22 case reports is included. A summary of the chapter contents is presented in Table One.

### Table One: References and Case Reports in Chapter Thirteen †

<table>
<thead>
<tr>
<th>Disease/Disorder</th>
<th>References (number)</th>
<th>Cases (number)</th>
<th>Cases (%)</th>
</tr>
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<tr>
<td>Hypertensive Disease</td>
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</tr>
<tr>
<td>Ischemic Heart Disease</td>
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<tr>
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<td>7</td>
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</tr>
<tr>
<td>Arteriovenous Malformations</td>
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</tr>
<tr>
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<td>6</td>
<td>3</td>
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</tr>
<tr>
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<tr>
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</tr>
<tr>
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<td>2</td>
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<tr>
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<td>Coagulation Defects</td>
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<tr>
<td>Other Blood Diseases</td>
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<tr>
<td>Totals</td>
<td>65</td>
<td>22</td>
<td>18.3%</td>
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</tbody>
</table>

† Total number of case reports in Part Two is 120.

* The International Classification of Diseases 9th Revision (ICD-9-CM) is a volume that provides an international standard for the classification of diseases. It was prepared by the Commission on Professional and Hospital Activities [Ann Arbor, Michigan: Edwards Brothers, Inc.], April 1986.
In 1981, this 57-year-old woman underwent placement of a sequential aortocoronary bypass graft to the right posterior descending, posterior circumflex marginal, middle circumflex marginal, and left anterior descending coronary arteries. She presented in 1983 with an acute anterior myocardial infarction, and at cardiac catheterization, the graft was totally occluded after its anastomosis to the posterior descending. Intracoronary thrombolysis was attempted but was unsuccessful. Right bundle branch block with left axis deviation developed, and a temporary transvenous pacemaker was placed in the right ventricular apex. The patient was then admitted to the coronary care unit for post-myocardial infarction care. Ventricular fibrillation occurred and was successfully treated. The next day, acute hypotension followed by electromechanical dissociation occurred and persisted for 45 minutes despite aggressive cardiopulmonary resuscitation. Finally, after intracardiac injections of calcium chloride and epinephrine were given as a last resort, acceptable hemodynamics developed. The patient awoke and quickly returned to her normal neurologic baseline, but dopamine support (20 micro/kg/minute) was required to maintain systolic blood pressure levels of 80 to 90 mmHg. Electrocardiography demonstrated an extensive anterior myocardial infarction.

The patient remained dopamine-dependent, with a pulmonary capillary wedge pressure of 15 mmHg. Mental status was normal, but renal shutdown occurred. After three days of marked oliguria, the serum creatinine level was 9.5 mg/dl, and the blood urea nitrogen level was 66 mg/dl. Despite extensive discussion with the patient and her family, she declined peritoneal dialysis. She expressed dissatisfaction with her function capacity and did not want further aggressive therapy to interfere with her apparently grim prognosis.

The patient became progressively more confused and agitated as a result of worsening azotemia and disconnected all of her intravenous lines. Despite the abrupt termination of dopamine therapy, the patient’s systolic blood pressure remained at the 80 to 90 mmHg range. In accordance with the wishes of the patient and her family, she was transferred to a non-monitored room for supportive care. The patient remained essentially anuric for eight days and became unresponsive. During this period, there was no oral or parenteral intake. Small doses of morphine sulfate and hydroxyzine were given to relieve excessive muscle tremor. No laboratory values were monitored.

On the ninth day after myocardial infarction, the patient was noted to be incontinent of a small amount of urine, and on the 10th day, she awoke and was capable of somewhat confused but alert conversation. Electrocardiography demonstrated an extensive anterior myocardial infarction.

The patient was discharged on the 26th hospital day. The blood urea nitrogen value was 33 mg/dl, creatinine value was 2.3 mg/dl, and serum electrolyte values were normal. Her systolic blood pressure was in the range of 90 to 100 mmHg. The radionuclide left ventricular ejection fraction was 40%. She noted significant easy fatigability and dyspnea on exertion. Angina was controlled with 60 milligrams of propranolol and 1 inch of nitroglycerin ointment every six hours.

Follow-up outpatient evaluation has been marked by slow improvement in exercise tolerance and a return of the patient’s renal function to pre-hospitalization levels, with a blood urea nitrogen value of 25 mg/dl and creatinine of 1.8 mg/dl.
Ischemic Heart Disease

Long-Term Survival in Cardiac Patients

HURWITZ MM; WHITE PD; EDWARDS JE
Geriatrics 24(2): Feb 1969; 86-96

Extracted Summary

A number of case histories are presented to illustrate the long survival of some cardiac patients. All physicians encounter such patients and, because of this ability to survive for many years, these patients are often advanced in age. Many examples of this type can be found in the literature. Long survival is possible in cardiac patients of all types, even some with congenital defects, and because of this, heart disease should be treated optimistically and without regard to the fact that the patient may be an individual of advanced years with heart disease of long standing.

Selected Case Report

Case 2: Angina pectoris for a few weeks at the age of 55, completely cleared by rest with excellent health eleven years later. F. D. was first seen by one of us (P. D. W.) in 1947 at the age of 47. He had always been very healthy, active, and busy and played a considerable amount of golf each summer. He had no symptoms but, since many of his friends had died recently of heart trouble, he became concerned about himself and desired a checkup. He smoked two or three cigarettes a day and rarely used alcohol. Results of the physical examination were entirely normal. His weight was 168 pounds. His pulse was regular at 60 and his blood pressure was 120 mmHg systolic and 80 mmHg diastolic. Fluoroscopy of the heart and lungs was normal as was the electrocardiogram. He was reassured and advised to continue full exercise. In May, 1951, he returned for a four-year check-up, feeling perfectly well, with no symptoms.

In early January, 1955, he thought he should have a checkup because of “slight indigestion recently.” He reported that he had continued to be very healthy and active until four weeks before the examination when, a few days after he stopped smoking and while very tired from overwork, he was troubled by substernal burning and oppressive pain which lasted a few minutes if he was walking far, fast, or upgrade. There was no radiation of the pain. This discomfort had recurring almost daily but never while he was at rest. He had experienced pain while playing ping pong and sawing wood. A recent gastrointestinal x-ray study had shown no abnormalities except for a questionable hiatus hernia.

Physical examination revealed that the heart sounds were good and there were no murmurs. The pulse was regular at a rate of 72. The blood pressure was 125 millimeters systolic and 80 millimeters diastolic. Fluoroscopy showed normal heart size and clear lungs. The electrocardiogram showed normal rhythm, rate 70, with low T1, inverted T waves in V2, V3, V4, and V5, suggesting coronary insufficiency in the anterior wall of the left ventricle. He was advised to rest at home for two weeks, eat a light diet especially low in animal fat, lose weight, and use nitrites whenever necessary.

On February 14, 1955, he reported that he had been very well during the previous four and 1/2 weeks with no need for nitroglycerine. Physical examination showed him to be looking well. His weight was 153 1/2 pounds, down 14 1/2 pounds. The pulse was regular at 54 and the blood pressure was 125 mmHg systolic and 80 mmHg diastolic. The heart examination showed good sounds, the first doubled at the apex, with no murmurs. The electrocardiogram was considerably improved but not yet normal. The serum cholesterol was 248 mg/100 ml.

He was re-examined at six-month intervals and by August, 1956, his electrocardiogram had returned to normal and he had no symptoms. When he was last seen in 1966 at the age of 66, his pulse was regular at 58, the blood pressure was 110 mmHg systolic and 70 mmHg diastolic and the heart sounds were good, the first doubled, with no murmurs. Fluoroscopy was normal. The electrocardiogram was normal in all 12 leads. He was considered to be in excellent health with complete recovery from his trouble eleven years earlier.

Comment: This patient illustrated several important points: the value of serial checkups to establish a base line of normality; the demonstration of the onset of temporary coronary heart disease by both symptoms and electrocardiogram; the complete clearing of the trouble, as shown by disappearance of both the angina pectoris and the electrocardiographic abnormality in the course of eighteen months, and the probable benefit of common sense measures such as weight loss, reasonable avoidance of stressful situations, temporary rest at home, and the liberal use of glyceryl trinitrate, without the imposition of complete invalidism.
Noninvasive and Invasive Demonstration of Spontaneous Regression of Coronary Heart Disease

ROTH D; KOSTUK WJ

Extracted Summary

Spontaneous regression of a left anterior descending coronary artery lesion was diagnosed by noninvasive testing (stress electrocardiography and thallium-201 myocardial imaging) and confirmed on selective coronary angiography in a 46-year-old man. The patient's clinical improvement, normalization of stress ECG and thallium-201 imaging, together with the loss of collateral filling, confirm that the regression is genuine. This case provides evidence that regression of coronary atherosclerosis can occur in man.

SELECTED CASE REPORT

A 46-year-old white male lawyer was admitted to the Cardiac Care Unit of University Hospital in July, 1977 after an episode of acute dyspnea, chest tightness and presyncope that started while he was playing soccer with his children. In the previous 8 months he had been aware of exertional chest pain that had increased in severity during the few months before his admission. One year earlier he had been told by his family physician that he had a slight increase of his serum triglyceride and cholesterol levels. He had stopped smoking in 1975, but before that had smoked one pack of cigarettes per day for 25 years.

His father had died at age 51 years from a myocardial infarction and a sister had died in her 40s from complications of diabetes mellitus. The functional inquiry was noncontributory.

Physical examination demonstrated a normal 46-year-old male in no distress, with a blood pressure of 120/80 mmHg and a pulse of 76 beats/minute. The only physical findings were the presence of arcus senilis, an S4 gallop and a soft apical systolic murmur. The remainder of the physical examination was unremarkable.

A 12-lead ECG was normal and remained unchanged over the next 5 days. The serum enzymes remained normal. The serum electrolytes, BUN and creatinine were normal. The serum uric acid and fasting glucose were 6.4 and 98 mg/dl respectively and the fasting serum cholesterol and triglycerides were 269 and 205 mg/dl, respectively. A chest x-ray was normal. Several days after admission, an exercise stress test was performed in conjunction with thallium myocardial imaging. The exercise test was discontinued at the end of the second stage (6 minutes) as the patient was experiencing pain that radiated into the left arm and ST depression. His heart rate was 150 beats/minute and the 12-lead ECGs showed up to 50 millimeters of downsloping ST-segment depression in the inferolateral leads. The thallium-201 myocardial images showed evidence of an anterior perfusion defect.

The patient was discharged on β blockade in the form of timolol, 5 milligrams three times daily. He discontinued this medication on his own accord within a few days because of side effects.

One month later coronary angiography was performed to assess the severity of the coronary atherosclerosis. The left ventricle demonstrated normal contractility, with a pressure of 125/12 mmHg and aortic pressure of 115/70 mmHg. A nondominant right coronary artery supplied collaterals that filled the left anterior descending coronary artery retrogradely. The proximal left anterior descending coronary artery had a significant stenosis at the origin of the first septal perforator which persisted even after the use of sublingual nitroglycerin. Surgery was advised on the basis of the patient's age and position of the left anterior descending coronary artery stenosis. However, the patient elected for a nonsurgical approach after the pros and cons of aortocoronary bypass were explained. During the ensuing 6 months the patient became more physically active and made a few changes in diet and life style, eating less meat and dairy products and reducing his law practice. Six months after his initial clinical presentation, his exertional chest pain resolved totally.

In May, 1978 the exercise test and thallium myocardial imaging were repeated. This time, the test was terminated at the end of the fourth stage (12 minutes of exercise), when the predicted maximal heart rate was reached without symptoms. Both the 12-lead ECG and the thallium scan were normal. A repeat coronary angiogram was obtained a few weeks later. The collateral filling of the left anterior descending coronary artery from the right coronary artery was no longer evident. The proximal lesion of the left anterior descending coronary artery was smaller. The fasting serum cholesterol and triglyceride values were 201 and 93 mg/dl, respectively.

The patient remains asymptomatic and his exercise ECG and thallium-201 myocardial scans are normal (September, 1979).
Spontaneous Remission is a Frequent Outcome of Variant Angina

WATERS DD; BOUCHARD A; THÉROUX P


Extracted Summary

To assess the prevalence of spontaneous remission in variant angina, 100 patients with this diagnosis who had undergone coronary arteriography in the hospital and a follow-up of at least 1 year were studied. Patients with coronary bypass surgery or myocardial infarction were excluded. Remission was diagnosed in 45 of the 100 patients who had been angina-free and had had no treatment for more than 3 months (mean 18.3). The other 55 patients were receiving medical treatment; 37 had been angina-free for at least 6 months (mean 22.5) and angina persisted in 18. The persistent-angina group had a lower prevalence of organic coronary stenoses 70% or greater: 4 of 18 versus 22 of 45 and 22 of 37 for the other two groups (p < 0.05), and a longer history of rest angina before admission. The remission group contained more patients (17 of 45 versus 4 of 55 [p < 0.001]) whose attacks had been documented only by provocative testing. Rest angina recurred when calcium antagonist drugs were discontinued in 15 of 51 instances, within 1 month in 11 patients and later in 4 patients. Remission was eventually attained in 35 of the 38 patients in whom these drugs were stopped. These results indicate that remission is a frequent outcome of variant angina. This fact should be considered in the evaluation of the long-term results of treatment and in the planning of care for an individual patient.

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Cerebrovascular Disease

ARTERIOVENOUS MALFORMATIONS

Spontaneous Disappearance of Cerebral Arteriovenous Angioma

Cas Report

CONFORTI P


*Extracted Summary*

The author reports the case of a 37-year-old woman in which a cerebral arteriovenous malformation disappeared completely and spontaneously within 3 years. Comparable cases from the literature are cited. The disappearance of the malformation may have been due to hemorrhagic episodes and the resulting thromboses. (Permission to reproduce case report denied by publisher.)

Spontaneous Closure of a Dural Arteriovenous Malformation

MAGIDSON MA; WEINBERG PE

*Surgical Neurology* 6: Aug 1976; 107-110

*Extracted Summary*

A case in which closure of an arteriovenous malformation involving the dura mater occurred spontaneously is presented. The anatomy, pathophysiology and clinical symptomatology of such lesions are reviewed. Possible mechanisms of spontaneous closure of intracranial arteriovenous malformations are discussed.

Spontaneous Regression of an Extra- and Intracranial Arteriovenous Malformation

Cas Report

HANSEN JH; SOGAARD I

*Journal of Neurosurgery* 45(3): Sept 1976; 338-341

*Extracted Summary*

The authors report a case in which an arteriovenous aneurysm located partly extracranially and partly in the posterior cranial fossa disappeared within 15 months without bleeding episodes or surgical intervention. The patient’s clinical symptoms, headache, and cranial bruit disappeared completely over the same period. (Permission to reproduce case report denied by publisher.)
A 28-year-old man was admitted on January 13, 1968, with a three-month history of a pulse-synchronous buzzing sound in the right ear. He had noticed it five months after a severe head injury with unconsciousness of a week's duration. The sound persisted for three months without changes in character or intensity.

On admission, the patient's physical and neurological examinations were normal as were his laboratory data. A loud pulsatile bruit was heard over the right mastoid region. The bruit disappeared immediately on compression of the right common carotid and the right occipital arteries. The right occipital artery pulsated more forcefully than the left occipital artery. Hearing and vestibular functions were found to be normal.

X-ray films of the skull revealed an irregular linear fracture close to the groove of the transverse sinus on the right side. The right common carotid angiogram and the right retrograde brachial angiogram demonstrated the transverse-sigmoid sinus, the superior petrosal sinus and the internal jugular vein in the arterial phase. Near the sinuses, a network of abnormal tortuous fine vessels was opacified. It was fed by the meningeal and the mastoid branches of the occipital artery, the posterior branches of the middle meningeal artery and the posterior meningeal branches of the vertebral artery. The tentorial artery from the internal carotid artery was faintly opacified. It supplied blood to the lesion. The right transverse sinus filled densely in the arterial phase. It was reduced in size and irregular in part, with small filling defects.

Ligation and excision of the right occipital artery was performed on March 7, 1968. This resulted in a slight decrease of the bruit. Several days after this operation the bruit returned as intense as ever. The postoperative right carotid angiogram revealed, in the early arterial phase, a dural arteriovenous malformation in the mastoid region just as in the previous angiograms. Various feeding arteries appeared larger and increased in number, except for the distal portion of the occipital artery. In the late arterial phase, the transverse-sigmoid sinus was outlined densely, accompanied by abnormal tortuous vascular nets near the sinus.

The patient rejected further surgical treatment and was discharged on April 10, 1968. During the ensuing nine years, he had no complaints other than the pulsatile tinnitus becoming faint and disappearing for a few days for no obvious reason. Physical and neurological examinations at this time revealed no abnormali-
ties. Auscultation over the right mastoid region failed to disclose any bruit. The right carotid angiogram, carried out in September, 1977, demonstrated no abnormalities and the vertebral angiogram proved to be normal. The right transverse sinus was not visible in either the carotid or the vertebral angiogram and was assumed to be occluded. There were no abnormal findings in hematologic tests or other blood survey. He had no complaints the following year.

Spontaneous Regression of Posterior Fossa Dural Arteriovenous Malformation

ENDO S; KOSHU K; SUZUKI J

*Journal of Neurosurgery* 51(5): Nov 1979; 715-717

**Extracted Summary**

The authors report a case of infratentorial arteriovenous malformation that regressed spontaneously within 25 months without bleeding episodes or surgical intervention. The patient's clinical symptoms, left-sided tinnitus, cranial bruit, and right homonymous hemianopsia disappeared completely over the same period. (Permission to reproduce case report denied by publisher.)

Long-Term Follow-Up Study by Cerebral Angiography in Cases with Arteriovenous Malformation of the Brain

with **Special Reference to Spontaneous Disappearance of Arteriovenous Malformation in Cerebral Angiography**

NUKUI H; MIYAGI O; TAMADA J; MITSUKA S; KAWAFUCHI J-I

*Neurologia Medico-Chirurgica* 22(2): Feb 1982; 125-132

**Extracted Summary**

Follow-up studies by cerebral angiography 4 to 11 years after the first angiography were carried out in six out of 44 cases with arteriovenous malformation of the brain (AVM) which were not operated on radically for various reasons. The size of the AVM was unchanged in five cases and spontaneous complete regression of the AVM was noted in one case. This 44-year-old man had a sudden attack of severe headache followed by disturbance of consciousness in April 1968. Bloody CSF was noted in spinal taps. He was admitted in August 1968. Vertebral angiography on admission showed an AVM of 1.4 x 1.1 x 1.0 centimeters in size in the left occipital lobe. The main feeder was the posterior cerebral artery and the drainer joined the vein of Galen. Craniotomy was performed, but no attempt was made to remove the AVM. Right hemiparesis and hemihypesthesia suddenly appeared and continued for 1 to 2 months four times from 1970 to 1978, but computerized tomography revealed no abnormal findings. The second angiography performed 10 years after the first study showed complete disappearance of the AVM. The same attack was noted in June 1979 and the third study revealed reappearance of a small AVM.

Spontaneous disappearance of the AVM was also demonstrated in another case during admission. This 26-year-old man had a sudden attack of left hemiparesis in January 1966. Right carotid angiography revealed a small AVM of 0.7 x 0.5 x 0.5 centimeters in size in the right parietal lobe. The main feeder was the pericallosal artery and the drainer joined the superior sagittal sinus. A small intracerebral hematoma was noted posterior to the AVM. He was treated conservatively. Angiography performed 3 months after the first study demonstrated complete disappearance of the AVM. The left hemiparesis recovered gradually and he returned to his job. No more attacks were noted for 7 years after discharge.
A 50-year-old laborer presented with a 48-hour history of headache of abrupt onset, vomiting, and a pulsating noise in the left side of his head. Clinical examination revealed no focal neurological abnormality in an alert patient. He had moderate neck stiffness, and a loud bruit could be heard over the left temporal region. The bruit could be reduced by ipsilateral carotid compression in the neck, but contralateral compression did not alter its character. His left retinal veins appeared slightly full and questionably arterialized.

On the lateral projection, skull x-ray films showed a curvilinear fracture involving the left parietal bone. (The patient later recalled minor head trauma a few years previously with no clinical symptoms nor radiological investigation.) Cerebral angiography was performed. There was elevation of the M-1 segment of the left middle cerebral artery and medial and superior displacement of the sylvian branches. A selective left external carotid angiogram showed a vascular malformation along the greater wing of the sphenoid bone, which was supplied primarily by the middle meningeal artery and which drained into the pterygoid plexus. There was no shift of midline structures. The right carotid angiogram and vertebral angiogram were normal. The studies were interpreted as showing a left sphenoparietal dural AVM with a middle fossa avascular mass lesion thought to represent an epidural hematoma.

Because there was no neurological deficit and the patient’s clinical condition was stable, he was managed conservatively. A cranial computed tomographic (CT) scan obtained 5 days later showed a small left anterior temporal lobe hematoma with surrounding edema, no surface clot, and no shift of the midline structures. The patient was discharged home 2 weeks after admission.

Four months later, repeat angiography was performed. Selective left internal and external carotid studies demonstrated no vascular malformation nor any other intracranial abnormality. The patient was free of symptoms and has remained so for the 18 months since.

**Extracted Summary**

The case of a 50-year-old man with a supratentorial dural arteriovenous malformation (AVM) associated with intracerebral hemorrhage is reported. Angiographically confirmed spontaneous regression of the AVM occurred without any form of surgical intervention. A possible mechanism of spontaneous closure of the AVM is offered.

**SELECTED CASE REPORT**

A cerebral arteriovenous malformation (AVM) spontaneously and completely disappeared on subsequent angiography. Computed tomography revealed a very similar picture to that of...
A 14-year-old boy experienced sudden severe headache, dizziness, and vomiting on February 28, 1977, and was admitted five days later. He was alert and complained of severe headache. There was neck stiffness without any focal neurologic deficit. No cranial bruit was audible. Blood pressure was 130/80 mmHg, pulse rate, 60 beats/minute. The remainder of the examination showed no abnormalities. Examination of the blood and urine also showed nothing abnormal. Lumbar puncture yielded xanthochromic CSF with normal opening pressure. Roentgenograms of the skull showed no calcification and no enlarged vascular grooves. The patient was discharged in one week.

There were several major motor seizures during the next three years. A EEG taken on March 16, 1979, was interpreted as moderately abnormal, demonstrating mild diffuse slowing of background activity and multiple spike discharges in the right frontal region.

The patient again experienced severe headache, nausea, and vomiting on March 30, 1980. He was admitted two days later. Neurologic examination showed only neck stiffness. Lumbar puncture disclosed bloody CSF with normal pressure. Routine laboratory findings were within normal limits.

Right carotid angiogram on April 6, 1980, disclosed an AVM in the basal portion of the right frontal lobe. This AVM was 4 x 4 x 3.5 centimeters in size, and was fed by the medial lenticulostriate arteries of the right middle cerebral artery and was drained by the sphenoparietal sinus. Excision was not attempted because the lesion was deep and quite extensive. He was discharged on April 7, 1980. After discharge, he did well.

The patient requested reevaluation of the AVM to provide a basis for promising new therapy. He was admitted on October 27, 1981, and showed no neurologic deficit. Bilateral carotid and left vertebral angiograms the next day disclosed complete disappearance of the AVM. A CT scan without infusion of contrast material showed an irregular, high-density mass in the right frontal lobe. The EMI (Electrical and Music Industries) number of this mass was between 75 and 173. The mass had no definite contrast enhancement. An EEG was interpreted as slightly abnormal, demonstrating increased theta waves over both temporal areas without spike discharge. Another CT scan taken six months later showed the same features.

He is now doing well. There has been no seizure for two years. He is taking 40 milligrams of phenobarbital sodium, 100 milligrams of phenytoin sodium and 200 milligrams of carbamazepine daily.

Complete Spontaneous Regression of a Cerebral Arteriovenous Malformation

LERAMO OB; CLARKE WFB

Southern Medical Journal 80(4): Apr 1987; 529-532

Extracted Summary

Complete spontaneous resolution of an intracranial AVM without any antecedent history of direct surgical intervention is uncommon. A review of the available literature yielded only seven cases, most of them in single case reports. Although spontaneous resolution is acknowledged as a rare feature in the complex natural history of AVM, its true incidence is unknown. Causes or factors that predispose to spontaneous regression of AVMs remain poorly understood. The majority of the documented cases of spontaneous resolutions have occurred in small or medium sized supratentorial lesions supplied predominantly by one major feeder. We report herein the unique occurrence of complete spontaneous regression of a deep left temporal AVM in a 22-year-old right-handed man. This case typifies the common picture, a clinical presentation of intracranial hemorrhage with raised intracranial pressure followed by complete resolution of the lesion.

Selected Case Report

This 22-year-old right-handed man was admitted in December, 1982 with a three-month history of severe headaches, visual obscuration, and intermittent diplopia. An ophthalmologist had diagnosed bilateral papilledema. The patient also gave a three-year history of grand mal seizures, which were usually preceded...
on each occasion by a vivid aura comprising a colorful visual hallucination of objects moving from the right visual field to the left, a funny smell “like burnt rubber,” and dysphasia. He had had five such episodes, the last occurring just two months before admission. Previous investigations in 1981, including a radionuclide brain scan, had shown no abnormality.

Clinical examination on admission was normal except for bilateral papilledema, which was worse in the left eye, and enlarged blind spots on perimetry. Skull x-ray films and EEG were normal. Brain scan showed an area of increased uptake in the left temporal region. Transfemoral carotid angiography showed an arteriovenous malformation and an avascular mass in the left temporal area with displacement of the left middle cerebral vessels superiorly and the left posterior cerebral artery medially. This AVM was supplied primarily by a hypertrophied left anterior choroidal artery together with some posterior choroidal vessels, and there was early filling of the basal vein of Rosenthal. Since there was no major neurologic deficit, and because the lesion was located deep in the dominant hemisphere, conservative treatment was elected. He was thus managed with anticonvulsants and dexamethasone with good results.

Follow-up assessment at six months in May, 1983 disclosed complete cessation of seizures and headaches as well as resolution of papilledema, with visual fields returning to normal. A repeat brain scan was normal. The left carotid angiogram done on May 26, 1983, and the vertebral angiogram on June 6 showed complete disappearance of the AVM. The branches of the middle cerebral artery had returned to their normal position and the anterior choroidal artery was of normal caliber. The supply from the posterior circuit had also disappeared.

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*Journal of Neurosurgery* 57(6): Dec 1982; 818-822

Spontaneous Regression of an Intrarenal Arteriovenous Malformation
YOSHIKI T; KONDO M; KITAYAMA T
*Journal of Urology* 137(4): Apr 1987; 725-726

Spontaneous Resolution of Arteriovenous Malformation Without Hemorrhage
MEGISON P; BATJER HH; PURDY PD; SAMSON DS
*American Journal of Neuroradiology* 10(1): Jan-Feb 1989; 204

**ANEURYSMS**

Spontaneous Resolution of an Intraventricular Hematoma
*Report of a Case with Recovery*

OJEMANN RG; NEW PFJ
*Journal of Neurosurgery* 20: 1963; 899-902

*Extracted Summary*

A case of intraventricular hematoma in the lateral ventricle, caused by rupture of a small arteriovenous malformation is reported. The location and size of the hematoma were documented by pneumoencephalography 7 weeks after the hemorrhage occurred. Spontaneous recovery of function to the prehemorrhage level occurred and a second air study 3 months after the onset of symptoms revealed virtually complete disappearance of the hematoma. The literature relevant to the problems of intraventricular hematoma is reviewed.
Spontaneous Thrombosis of Cerebral Aneurysms

LODIN H


Extracted Summary

The natural course of cerebral aneurysms, both those observed in subarachnoid haemorrhage and those discovered by chance, has been subject to much discussion. With a view to elucidating further the natural course of cerebral aneurysms, two patients with spontaneously thrombosed aneurysms are reported.

Selected Case Report


Spontaneous Remission of an Intraventricular Hemorrhage

Case Report

LOESER JD; STUNTZ JT; KELLY WA


Extracted Summary

We have presented a case of spontaneous intracerebral hemorrhage with intraventricular extension in a 55-year-old hypertensive Negro male. The intraventricular clot was originally demonstrated by pneumoencephalography, and spontaneous lysis was evident in a follow-up air study 2 months later. No operative therapy was undertaken and the patient's neurological status progressed from initial hemiplegia, hemihypesthesia and homonymous hemianopsia to mild paresis and sensory deficits 4 months after the ictus. The literature on intraventricular hemorrhage has been reviewed; this case and that of Ojemann and New contrast with the poor prognosis usually given to intraventricular extensions of spontaneous intracerebral hemorrhages. Progression of neurological signs or failure to improve, rather than simply the presence of an intraventricular clot, should determine the need for operative intervention. (Permission to reproduce case report denied by publisher.)

Spontaneous Cure of Intracavernous Aneurysm of the Internal Carotid Artery in a 14-Month-Old Child

Case Report

DEVADIGA KV; MATHAL KV; CHANDY J

Journal of Neurosurgery 30: Feb 1969; 165-168

Extracted Summary

We have reported the spontaneous thrombosis of an intracavernous aneurysm of the left internal carotid artery in a 14-month-old boy. Although sudden onset of ophthalmoplegia suggested a temporary carotid cavernous fistula, the abundant collateral blood supply to the brain even in the first angiogram indicated the probable congenital nature of the lesion. (Permission to reproduce case report denied by publisher.)
Spontaneous Cure of Intracranial Aneurysm

Case Report

KOWADA M; TAKAHASHI M; GITO Y
Acta Neurochirurgica 31: 1974; 131-137

Extracted Summary

A case of “spontaneous cure” of an intracranial aneurysm is reported and 7 cases verified by follow up angiography are reviewed. The autopsy finding of spontaneously thrombosed aneurysm is discussed.

Selected Case Report

A 42-year-old male entered our hospital with a chief complaint of high fever on August 11, 1971. He was treated for septicemia, because streptococcus viridans was found on blood culture. On the morning of September 11, he became stuporous soon after having complained of severe headache and vomiting. Lumbar puncture showed haemorrhagic cerebrospinal fluid with an initial pressure of 450 mm H₂O. The patient regained consciousness about five hours after this episode. Bilateral carotid and vertebral angiography was carried out by transfemoral catheter technique on November 13, and a saccular aneurysm 8 x 5 millimeters in size was demonstrated at the trifurcation of the left middle cerebral artery.

After he had recovered from his infection the patient was referred to the clinic of neurosurgery on February 2, 1972, for the radical treatment of his intracranial aneurysm. The operation was performed on February 7, and the trifurcation of the left middle cerebral artery was exposed. Surprisingly, the saccular aneurysm demonstrated on the angiogram was not found. A pouch-like elevation 4 x 3 millimeters in size, arising from the vessel itself, was seen at the trifurcation which was then wrapped with muscle. The previously revealed aneurysm was not demonstrated by angiography carried out the following day and again 2 weeks later. The patient was discharged on May 3, 1972.

Spontaneous Cure of Ruptured Intracranial Arterial Aneurysms

SPALLONE A; PERESEDOV VV; KANDEL EI
Surgical Neurology 16(5): Nov 1981; 367-370

Extracted Summary

The case of a patient who had spontaneous cure of an intracranial saccular aneurysm, documented by angiography, is reported. This occurred in a 41-year-old patient, admitted four months after recurrent subarachnoid hemorrhage due to an angiographically verified supraclinoid internal carotid artery aneurysm. The relevant literature is reviewed, and the possible mechanism of spontaneous aneurysmal thrombosis is briefly discussed. It is concluded that repeating angiography is not without merit in patients with already documented cerebral aneurysms who are referred for surgical treatment some time after a subarachnoid hemorrhage.

Selected Case Report

A 41-year-old, right-handed man was admitted in October 1980. In Jun, 1980, he had had an episode of sudden severe headache, followed for a few days by slight nuchal rigidity. This had prompted admission to a local hospital. He was referred to the local neurological department a few weeks later, shortly after a recurrent subarachnoid hemorrhage, verified by lumbar puncture. At admission the patient was alert but slightly disoriented. Neurological examination revealed nuchal rigidity, right third nerve palsy, slight left facial weakness, and increased deep tendon reflexes on the left. His blood pressure was 160/100. The patient had no previous history of hypertension. Lumbar puncture produced a hemorrhagic spinal fluid. A right carotid angiogram performed on the second day after the subarachnoid hemorrhage showed a supraclinoid aneurysm located at the origin of the posterior communicating artery.

The patient was treated conservatively with bed rest and antihypertensive drugs, and his neurological abnormalities cleared within a few days. He was later referred to
Part Two: Diseases Other Than Cancer

Circulatory System, Blood, and Blood Forming Organs

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Diseases of Arteries, Arterioles and Capillaries

Polyarteritis Nodosa: Chronic Renal Failure with Spontaneous Recovery

Szwed JJ; Gaither JM; Kesler PA

American Journal of the Medical Sciences 286(2): Sep-Oct 1983; 36-40

Extracted Summary

A 57-year-old male developed acute oliguric renal failure due to polyarteritis nodosa (PAN). He went on rapidly to chronic renal failure. After nine months of home hemodialysis, he recovered renal function with creatinine clearance of 30 ml/minute. This patient was not treated with drugs of any type for his illness. He represents a spontaneous remission from severe renal PAN and underscores the need for further evaluation of the current recommended drug therapy for this disease.

Selected Case Report

A 57-year-old man was admitted to his local hospital for severe ischemic necrosis of his fingers and papular dermatitis of his lower extremities which had evolved over the two months prior to admission. At the time of his initial hospitalization, he had a normal blood pressure, a mild anemia (hemoglobin 12.2), and a urinalysis with 1+ protein, 3+ blood, two to five WBC/high powered field and a few granular casts/low powered field. Westergren sedimentation rate was 49 mm/hour. The ANA, LE prep, and cryoglobulins were negative. Protein electrophoresis revealed decreased serum globulin. In his local hospital, the patient developed edema and hypertension, and noted reduced urine output and gross hematuria. Nephrologic and urologic history disclosed no previous disease.

On admission to Indiana University Medical Center three weeks later, the patient was found to be tachypneic and his physical examination showed a blood pressure of 140/90, a third heart sound, and dry gangrene of the right second and third fingers. Chest x-ray was normal, and the EKG revealed prolonged Q-T intervals. The serum sodium was 121 mEq/l, potassium 7.1 mEq/l, chloride 84 mEq/l and bicarbonate 14 mEq/l. BUN was 212 mg/100 ml, creatinine 17.2 mg/100 ml, calcium 7.7 mg/100 ml, and phosphate 12 mg/100 ml. Hemoglobin was 7.2 gm, hematocrit 22%. WBC was 7,800 with a normal differential and platelets were 202,000. Urinalysis revealed granular casts, white cell casts, 2+ protein, pH 6 and specific gravity was 1.008. Arterial blood gases revealed pO2 - 87, pCO2 - 25, and pH 7.37. Cryoglobulin, LE prep and ANA were negative. Quantitative immunoglobulins revealed mildly increased IgG; complement (C'3) levels were normal. Protein electrophoresis revealed reduced albumin and increased alpha, alpha 2 and beta globulins. Westergren sedimentation rates ranged from 65-112 mm/hour and eosinophil count was 357. Iron, as well as total iron binding capacity, were normal. Haptoglobin was 236 (normal up to 200). B12 and folate levels were normal. Blood cultures were negative. VDRL was non-reactive; HbsAg and monospot were negative. ASO was 240 Todd units. Bence-Jones proteins were positive with kappa and lambda chains both present. Bone marrow evaluation revealed a normal stimulated marrow; nasal septal biopsy revealed nonspecific inflammation. Renal biopsy revealed crescentic glomerulonephritis in 80% of the glomeruli and most glomeruli (45/50), 90%, with complete obliteration and destruction of the capillary tuft. IgA, C'2, IgG, and C'4 staining was negative. An admixture of inflammatory cells including neutrophils, eosinophils, plasma cell and lymphocytes were present in the interstitial space.

our clinic for possible stereotactic clipping. On admission, general and neurological examinations were unremarkable. The blood pressure was 130/75. A left carotid angiogram was performed in order to exclude contralateral abnormalities and to test the collateral circulation. It was negative. Right carotid angiography, performed a few days later, failed to reveal any pathological change despite repeated injections of contrast material. The patient was subsequently discharged. Four months later he was still free of complaints.
These infiltrates were present particularly around necrotic glomeruli and arteries in which fibrin thrombi and fibrinoid necrosis could be demonstrated. These histologic findings were consistent with acute polyarteritis nodosa. Renal angiography revealed changes consistent with necrotizing vasculitis of the kidney including multiple aneurysm formation.

Urine output was averaging 250 ml/24 hours and after 2 weeks in the University Hospital, there was no evidence of improvement in renal function. The patient was hemodialyzed via an arteriovenous shunt during this early period and later converted to an arteriovenous fistula for home dialysis. No drug therapy of any type was administered. That is, he received no steroids nor immunosuppressives for his polyarteritis nodosa at any time.

The patient was discharged to begin home dialysis training after four weeks. He successfully mastered home dialysis and was discharged to be followed every month in outpatient clinic. The ischemic lesions of his hands and feet resolved within 2 months of discharge. Nine months after initiation of dialysis, the patient has been found to have a creatinine clearance of 30 ml/minute with a plasma creatinine of 2.8 mg/100 ml and he has been advised to discontinue dialysis entirely. A urinalysis revealed a specific gravity of 1.005, pH of 5, 2+ protein and occasional granular casts/low powered field. His blood pressure was 140/80. Presently he is regularly employed and asymptomatic on no drug therapy. He has been in remission for eleven months.

### Spontaneous Regression of a Dura Mater Arteriovenous Fistula of the Posterior Fossa

**ROHR J; GAUTHIER G**  
*Revue Neurologique 141(3): 1985; 240-244*

**Extracted Summary**

A case of spontaneous regression of a posterior fossa dural arteriovenous malformation supplied by the left external carotid artery is reported. The patient, a 68-year-old woman, complained of pulsatile tinnitus most importantly at night. Two years after the malformation discovery, she presented a transient ischemic attack in the left middle cerebral artery territory. A new angiography showed that the malformation had disappeared; the left transverse sinus could not be opacified. The authors emphasize an interest in the use of Doppler sonography for detection and follow-up of such malformations. They review the few published similar cases and discuss the different possible closure mechanisms of intracranial arteriovenous malformations.

### Supplemental References

**Diseases of Arteries, Arterioles, and Capillaries**

- **Spontaneous Disappearance of Iatrogenic Renal Arteriovenous Fistula: Report of a Case**  
  **HERSCMAN A; KLEIN MJ; BLUMBERG AG**  
  *Journal of Urology 105(1): Jan 1971; 4-6*

- **The Problem of Spontaneous Healing of Carotid-Cavernous Fistulae (Zur Frage der Spontanheilung der Karotis-Kavernosus-Fistel)**  
  **GRUNERT V; SUNDER-PLASSMANN M; VALENCAK E**  
  *Fortschritte auf dem Gebiete der Roentgenstrahlen 117(5): Nov 1972; 548-551*

- **Spontaneous Resolution of a Descending Aortic Dissection. Report of a Case**  
  **THORVINGER B; ALBRECHTSSON U**  
  *Acta Radiologica 30(3): May-Jun 1989; 305-306*
Spontaneous Regression of Budd-Chiari Syndrome (Hepatic Venous Occlusion) in a Young Female

Hultcrantz R; Angelin B; Einarsson K; Friman L
Acta Medica Scandinavica 221(5): 1987; 503-507

Extracted Summary

A case of occlusion of the hepatic veins in an 18-year-old girl is presented. The onset was sudden with massive ascites and markedly impaired general condition. The diagnosis was based on liver biopsy and angiograms of the caval and hepatic veins as well as of the celiac artery. No predisposing factors could be found. The patient was treated conservatively with laparocentesis and diuretics. Clear improvement was seen after two weeks, and after four weeks she had no ascites and could be discharged. All liver function tests were then normalized. After three months, all diuretics could be withdrawn, and in the following 11 years she had remained completely recovered. The case illustrates also that widespread thrombi of the hepatic veins may sometimes rapidly dissolve spontaneously, with apparent total reconstitution of hepatic function. This case is unusual since previously reported cases have had high mortality rates and, in surviving cases, operative procedures or large doses of diuretics have been required to control ascites.

Selected Case Report

The patient was an 18-year-old, previously healthy Swedish girl without a history of drug treatment including contraceptive steroids. Two weeks before admission she had right-sided abdominal pain, and a right-sided ureteral concrement was diagnosed by an i. v. pyelogram. Following spasmolytic treatment she was relieved of symptoms, and a repeat pyelogram was normal. Three days before admission she noticed swelling of the abdomen, and when this progressed she was seen at the emergency room. On examination she had extensive ascites and splenomegaly, but the liver was not palpable. A gynecological examination was normal. After unsuccessful laparoscopy, a laparotomy was carried out on the next day because of suspicion of malignancy, and 4.5 liters of sanguinolent ascites were removed. The liver was coloured red and congested. The spleen was twice the normal size. The portal pressure, measured during the laparotomy, was 47 centimeters H2O. A needle biopsy from the right liver lobe showed acute congestion around the central veins and the surrounding parts of the sinusoids. No signs of cirrhosis or inflammation could be seen. Around some central veins parenchymal cell necrosis was noted. A gastroscopy two days later showed small varices in the distal oesophagus. After six days of postoperative care, the patient was transferred to the Department of Internal Medicine at Serafimer Hospital for further investigation. On admission the patient still had massive ascites, tachypnea, and felt weak. She had evidence of right-sided pleural effusion.

Laboratory findings: A slight anemia (Hb 100 gm/l) was present, while the white cell count and thrombocytes were normal. Serum transaminases were slightly elevated and alkaline phosphatases were raised to twice normal, while bilirubin and creatinine were normal. The prothrombin time was slightly prolonged. Albumin was 40 gm/l, and a routine plasma electrophoresis showed a normal pattern. Values for alpha, antitrypsin and ceruloplasmin were normal. Bromsulphalein retention after 45 minutes was 10%. The bone marrow did not show any pathological features besides reactive changes and lack of iron. An extensive investigation ruled out the possibility of a coagulopathy. Antibody titers towards hepatitis B, smooth muscle cells, mitochondria, and DNA were not found. There was no evidence of CMV or mononucleosis infection. Investigations for echinococcus or bilharzia were negative. ECG and phonocardiogram were normal.

Radiology: A chest x-ray was normal except for a small amount of fluid in the right pleura. Liver spleen scintigraphy showed enlargement of the left liver lobe and the spleen, but no increase in bone marrow activity. Angiography of the inferior caval vein demonstrated narrowing of its intrahepatic part and compression from the left. Hepatic venography failed to demonstrate any normal hepatic veins, but strange-looking veins, which had to be interpreted as collaterals, were filled from an area of parenchymal opacification. The maximum filling of the portal vein at celiac angiography occurred late, indicating a decreased portal flow. The right liver lobe was compressed by ascites and the arteries in the right lobe were somewhat tortuous, but the arteries in the left lobe were...
stretched. The wedged hepatic venous pressure was 37 centimeters H₂O.

Clinical course: After initial laparocentesis during which 6 liters of ascites were withdrawn, the ascites could be controlled with orally given diuretics (160 milligrams furosemide and 100 milligrams spironolactone daily). The production of ascites seemed to decrease and diuretics could be gradually reduced. After four weeks the patient was fully recovered and could be discharged. After three months all diuretics had been withdrawn without any complications and all liver function tests were normal. A liver-spleen scintigraphy one year after the onset of the disease still showed a slight enlargement of the left liver lobe, but was otherwise normal. During a follow-up of 11 years the patient has remained in full health without clinical or laboratory evidence of liver disease.

Congenital Anomalies

Atrial Septal Defects

Spontaneous Closure of an Atrial Septal Defect

TIMMIS GC; GORDON S; REED JO

*Journal of the American Medical Association* 196(1): April 4 1966; 137-139

Extracted Summary

Serial hemodynamic and angiocardiographic studies demonstrated the spontaneous closure of an atrial septal defect in a 3-year-9-month-old white boy, the presence of which was confirmed in the second year of life. A possible myocarditis in the first year of life was considered to be potentially instrumental in the closure of this defect. Although similar observations have been made by others, to our knowledge this is the first such case, particularly at this age, reported in the literature.

Spontaneous Functional Closure of Symptomatic Atrial Septal Defects

CAYLER GG


Extracted Summary

Spontaneous functional closure of large atrial septal defects was demonstrated by cardiac catheterization and cineangiographic technics in 3 infants. These 3 patients and 1 other with clinical evidence of spontaneous closure constitute 31% (4 of 13) of infants with acyanotic congenital heart disease and large secundum atrial septal defects studied at the Sutter Memorial Hospital in the past four years (1962-1966). Disappearance of large atrial left-to-right shunts was documented between the ages of seven days and fifteen months (Case 1), three weeks and eight months (Case 2) and three months and twenty months (Case 3). Case 1 had an associated moderate-sized ductus and aortic stenosis, Case 2 an associated small ductus, and Case 3 no other cardiovascular anomaly. The possible mechanisms of spontaneous closure are discussed.

The demonstration of spontaneous disappearance of large atrial left-to-right shunts supports a conservative approach to early surgical closure of atrial septal defects even in patients with large left-to-right shunts, cardiac enlargement and congestive heart failure. The spontaneous closure of atrial septal defects associated with complicated cyanotic cardiac anomalies has not been documented but probably occurs and obviously could have a profound effect on the natural history of such lesions.
Case 3: K. C., a 29-month-old girl, was admitted to Sutter Memorial Hospital at 3 months of age because of signs of progressive respiratory distress. A murmur had been noted 1 week previously. The birth weight was 3,260 grams (7 pounds, 3 ounces) and had risen to only 4,990 grams (11 pounds). Gestation and birth had been normal.

On physical examination an “elf-like” facies was noted. There was no cyanosis or clubbing. There was a bulge of the left side of the chest and a hyperdynamic parasternal cardiac impulse. No thrill was felt. The 1st sound was of increased intensity and maximal at the lower left border of the sternum. The 2d sound was of normal intensity and maximal at the upper left sternal border, and the split was increased and fixed. There was a Grade 3/6 systolic ejection murmur at the upper left sternal border. A 3d sound and a Grade 2/6 mid-diastolic rumble were present at the lower left border of the sternum. The liver edge was palpable 3 centimeters below the left, and the spleen edge 3 centimeters below the right costal margin. The peripheral pulsations were equal and of normal quality. There was no edema. A roentgenogram of the chest showed moderate cardiac enlargement and pulmonary plethora. There was right middle lobe atelectasis. An electrocardiogram disclosed right axis deviation (+150) and hypertrophy of the right ventricle. Digitalis (Lanoxin) was administered, and a low-salt formula (S-29) started.

Cardiac catheterization and cineangiography 1 week later indicated a large left-to-right shunt through a large atrial septal defect. There was minimal pulmonary artery hypertension and normal pulmonary vascular resistance.

The patient was discharged from the hospital 3 days after the catheterization and continued on digitalis and low-salt formula.

At 6 months of age the weight had risen to only 5,670 grams (12 pounds, 8 ounces). The cardiac physical findings, roentgenogram and electrocardiogram were essentially unchanged. By the age of 1 year she weighed 9.1 kilograms (20 pounds). There was no hepatosplenomegaly. She continued to have a deformity of the left half of the chest and a lifting parasternal cardiac impulse. The increased fixed split of the 2d sound persisted; however, the systolic murmur decreased to Grade 2/6, and the diastolic murmur disappeared. A roentgenogram showed only slight cardiac enlargement and slightly increased vascular markings. An electrocardiogram yielded less evidence of hypertrophy of the right ventricle.

At the age of 20 months she was readmitted to Sutter Memorial Hospital for recatheterization. She weighed 10.4 kilograms (23 pounds). There was no chest deformity. There was a normal apical cardiac impulse. The heart sounds were normal. There was a Grade 1/6 systolic ejection murmur at the upper left sternal border. No diastolic murmur was heard. The roentgenogram of the chest was now normal. The electrocardiogram showed right-axis deviation (+105), possible hypertrophy of the right ventricle and incomplete right-bundle-branch block.

Cardiac catheterization provided no evidence of left-to-right shunting by blood gas analysis or by the platinum electrode hydrogen method. At 29 months of age the patient had no cardiovascular symptoms and no murmurs.

### Functional Closure of Atrial Septal Defects

**CUMMING GR**

American Journal of Cardiology 22: Dec 1968; 888-892

**Extracted Summary**

Three patients with left to right shunts at the atrial level and clinical findings of atrial septal defect were studied at 8 months to 2 years of age. The subsequent clinical course of these children indicated that the atrial defect had closed. The second heart sound became normal although systolic ejection murmurs remained. Hemodynamic studies confirmed the functional closure of the atrial septal defect although probe patency of the foramen ovale was still present in 2 patients. Although functional closure of atrial septal defects may not be a common occurrence, the possibility should be kept in mind and elective surgery should not be undertaken during the first few years of life.

**Selected Case Report**

Case 1: This girl was admitted for cardiac studies at age 9 months. A murmur was noted at birth. Birth weight was 4.1 kilograms. At age 9 months weight was only 7.3 kilograms (tenth percentile), and the baby was not sitting or crawling. There were no specific cardiac symptoms. The cardiac impulse suggested right ventricular enlargement. The second heart sound was widely split and the split did not vary with respiration. There was a systolic ejection click and a grade 3/6 systolic murmur at the second left intercostal space. The electrocardiogram showed an rs'R' in lead V₆ with the R' wave measuring 23 millimeters. Chest roentgenogram showed a cardiothoracic ratio of 8/14 with some accentuation of lung markings. Heart catheterization studies confirmed the
A 6-year-old girl was referred to the Cleveland Clinic Foundation for elective surgical repair of an atrial septal defect. She was born ten weeks prematurely to an insulin-dependent diabetic mother, but had no perinatal problems. The patient had frequent respiratory tract infections when she was 5 to 6 months old, and at age 7 months she was seen by a physician because of a continuous "cold". During that examination a grade 2/6 systolic murmur was noted. An ECG showed right ventricular hypertrophy and a chest x-ray film demonstrated cardiomegaly with increased pulmonary vascularity. A cardiac catheterization was performed at that time. Saturation data revealed a step-up in saturation in the region of the low right atrium with equalization of right and left atrial mean and phasic pressures. The left atrium was entered via the interatrial communication. Injection of dye into the left atrium in the region of the left atrial appendage revealed a moderate-sized interatrial left-to-right shunt. The patient had been given digitalis and was followed up medically for six years. Growth and development were normal; however, she continued to have frequent respiratory tract infections. When she was referred to us for elective repair she was acyanotic, with a heart rate of 84 beats/minute and regular. When she was sitting the blood pressure was 95/50 mmHg in the right arm. There was some prominence of the left side of the chest. The lungs were clear. There was a right ventricular heave. The first heart sound was normal. The second was widely split but seemed to move normally with respiration. The pulmonary component of the second heart sound was normal. No murmur was noted. An ECG demonstrated a QRS axis in the frontal plane of +80 degrees with no evidence of hypertrophy. The chest x-ray film showed a normal-sized heart with a prominent main pulmonary artery but with normal pulmonary vascularity. A two-dimensional echocardiogram revealed a normal-sized right ventricle with no paradoxical septal motion. Contrast two-dimensional echocardiography with indocyanine green (Cardio-Green) revealed no right-to-left shunt at the atrial level. The subcostal view of the interatrial septum clearly demonstrated a small bulge in the mid region of the septum suggestive of a small interatrial septal aneurysm.

**Spontaneous Closure of Atrial Septal Defect with Interatrial Aneurysm Formation**

*Documentation by Noninvasive Studies, Including Digital Subtraction Angiography*

AWAN IH; RICE R; MOODIE DS

*Pediatric Cardiology* 3(2): 1982; 143-145

**Extracted Summary**

A 6-year-old girl had a well-documented atrial septal defect that underwent spontaneous closure with formation of an interatrial septal aneurysm. Closure was documented by noninvasive studies. To our knowledge, this is the first case of closure documented by digital subtraction angiographic technique.

**Selected Case Report**

A 6-year-old girl was referred to the Cleveland Clinic Foundation for elective surgical repair of an atrial septal defect. She was born ten weeks prematurely to an insulin-dependent diabetic mother, but had no perinatal problems. The patient had frequent respiratory tract infections when she was 5 to 6 months old, and at age 7 months she was seen by a physician because of a continuous "cold". During that examination a grade 2/6 systolic murmur was noted. An ECG showed right ventricular hypertrophy and a chest x-ray film demonstrated cardiomegaly with increased pulmonary vascularity. A cardiac catheterization was performed at that time. Saturation data revealed a step-up in saturation in the region of the low right atrium with equalization of right and left atrial mean and phasic pressures. The left atrium was entered via the interatrial communication. Injection of dye into the left atrium in the region of the left atrial appendage revealed a moderate-sized interatrial left-to-right shunt. The patient had been given digitalis and was followed up medically for six years. Growth and development were normal; however, she continued to have frequent respiratory tract infections. When she was referred to us for elective repair she was acyanotic, with a heart rate of 84 beats/minute and regular. When she was sitting the blood pressure was 95/50 mmHg in the right arm. There was some prominence of the left side of the chest. The lungs were clear. There was a right ventricular heave. The first heart sound was normal. The second was widely split but seemed to move normally with respiration. The pulmonary component of the second heart sound was normal. No murmur was noted. An ECG demonstrated a QRS axis in the frontal plane of +80 degrees with no evidence of hypertrophy. The chest x-ray film showed a normal-sized heart with a prominent main pulmonary artery but with normal pulmonary vascularity. A two-dimensional echocardiogram revealed a normal-sized right ventricle with no paradoxical septal motion. Contrast two-dimensional echocardiography with indocyanine green (Cardio-Green) revealed no right-to-left shunt at the atrial level. The subcostal view of the interatrial septum clearly demonstrated a small bulge in the mid region of the septum suggestive of a small interatrial septal aneurysm.

Cardiac nuclear pulmonary flow systemic flow ratio was normal with no evidence of any left-to-right shunt. The right ventricular ejection fraction was normal at 58% (normal for our laboratory ≥ 45%), and the left ventricular ejection fraction was 55% (normal for our laboratory, ≥ 55%).

Renografin-76 (diatrizoate sodium and diatrizoate meglumine) was injected into an antecubital vein at 10 ml/second for 2 seconds, using an 18-gauge angiocatheter. Digital subtraction angiography was performed with a DR-960 (Technicare Corporation) digital subtraction angiographic unit using the frontal and left anterior oblique 40° views. All images were made in the radiographic, single-mask mode and recorded on magnetic tape. A typical radiographic sequence utilized 80 to 100 kilovolt (peak), 5 to 10 mA per frame (2 to 10 milliseconds at...
1,000 mA), and up to six frames per second for 15 seconds. A single “best” mask was selected for immediate postexposure processing. The image display machine had 256 x 256 units, each with 256 shades of gray. The digital subtraction angiogram, particularly the left anterior oblique section, demonstrated no evidence of shunt at atrial level.

Supplemental References

Atrial Septal Defects

Serial Hemodynamic Observations in Secundum Atrial Septal Defect with Special Reference to Spontaneous Closure
Mody MR
*American Journal of Cardiology* 32: Dec 1973; 978-981

Spontaneous Closure of Secundum Atrial Septal Defect in Infants and Young Children
Cockerham JT; Martin TC; Gutierrez FR; Hartmann AF Jr; Goldring D; Strauss AW
*American Journal of Cardiology* 52(10): Dec 1 1983; 1267-1271

Ventricular Septal Defects

Spontaneous Closure of Ventricular Septal Defects
Evans JR; Rowe RD; Keith JD
*Circulation* 22: Dec 1960; 1044-1054

Extracted Summary

A group of 37 children is described in whom a systolic murmur heard early in life gradually diminished and eventually disappeared. When the patients were first seen, the clinical findings suggested a small ventricular septal defect but no thrill was present and the systolic murmur had a superficial blowing quality with high-frequency vibrations and tended to stop before the second heart sound.

Cardiac catheterization demonstrated a small left-to-right shunt at ventricular level in 4 of the patients while the murmur was present; in 1 this was repeated after the murmur had gone and no abnormality could be demonstrated. Cardiac catheterization in other patients with typical disappearing systolic murmurs showed a left-to-right shunt in some but in others this was too small to be detected by routine oxygen studies. A rough correlation was established between the length and intensity of the murmur and the size of the shunt. With angiocardiography and intracardiac phonocardiography the exact site of the ventricular septal defect was localized to the muscular portion of the septum in 4 of the patients.

In 1 patient who presented with congestive heart failure, clinical and hemodynamic findings of a large ventricular septal defect diminished over several years and finally disappeared.

Children with the specific type of systolic murmur described may be recognized as having a small defect in the muscular ventricular septum. The defect is thought to be gradually reduced in size and ultimately closed by hypertrophy of septal muscle. Spontaneous closure appears to be not uncommon with small ventricular septal defects and may rarely occur with lesions large enough to present with congestive heart failure.
Case 1: M. E. J., a 6-year-old female nonidentical twin, was delivered 3 weeks prematurely by cesarean section. The birth weight was 1.8 kilograms (4 pounds). A systolic murmur was heard at 5 weeks of age. Congestive heart failure was first diagnosed at 8 months of age on the basis of failure to thrive, irritability, dyspnea, hepatomegaly and cardiomegaly. The clinical diagnosis of a ventricular septal defect was made, but patent ductus arteriosus was also considered in the differential diagnosis. Oral administration of digoxin did not significantly ameliorate the symptoms.

At 15 months of age, on account of her unsatisfactory response to adequate doses of digitalis and the possibility of the presence of a patent ductus arteriosus, she was admitted to the Children's Hospital Medical Center for cardiac catheterization. Physical examination revealed the weight was below the 3d and the height below the 10th percentile on a standard developmental chart. The blood pressure was 95/55 over the right arm. The left side of the chest was prominent. A systolic thrill was maximal at the lower left sternal border. The cardiac impulse was hyperdynamic and involved the right and left ventricles. The 2d sound at the pulmonary area was normally split, and the pulmonic component was moderately accentuated. A 3d sound was present at the apex. A Grade 4 pansystolic murmur was heard at the lower left sternal border, and a Grade 2 mid diastolic rumble was noted at the apex. An electrocardiogram showed biventricular hypertrophy, and x-ray study revealed cardiac enlargement, with increased pulmonary vasculature. Cardiac catheterization revealed a left-to-right shunt at the ventricular level, with a pulmonary to systemic flow ratio of almost 3:1. The arterial blood was fully saturated, and the pulmonary arterial pressure was moderately elevated. No evidence suggesting the presence of a patent ductus arteriosus was found, and the patient was discharged from the hospital on continued digitalis therapy. She was followed in the outpatient department at 6-month intervals, during which she showed progressive improvement. Paralleling her more satisfactory weight gain, the disappearance of dyspnea and improved disposition, the systolic murmur gradually decreased in intensity.

At the age of 4 years and 7 months she was admitted to the hospital for recatheterization. At this time physical examination revealed that she was at the 25th percentile on the developmental chart by both height and weight. No chest deformity could be noted. Only a Grade 1, early short systolic murmur was heard at the left sternal border. This early systolic murmur could not be demonstrated phonocardiographically until a dose of ephedrine was administered, after which an early, short systolic murmur at the 4th left interspace appeared. In addition, an “ejection” type murmur was recorded at the 2d left interspace. An electrocardiogram was now normal for age. The radiogram showed slight cardiac enlargement, but the pulmonary vasculature was unremarkable.

Cardiac catheterization gave no evidence of a shunt, and the pulmonary-artery pressure was within normal limits. Cineangiograms, with an injection of dye into the left atrium, showed a puff of dye traversing the ventricular septum from left to right. This fifteen-month-old girl in congestive heart failure was shown at cardiac catheterization to have a large ventricular septal defect, with moderate pulmonary arterial hypertension. Repeat studies at four and a half years of age revealed virtual disappearance of the murmur, no left-to-right shunt detectable by oximetry and normal right ventricular pressures. Cineangiograms suggested the presence of a minute residual left-to-right shunt.
Ventricular Septal Defect

Incidence, Morbidity, and Mortality in Various Age Groups

KEITH JD; ROSE V; COLLINS G; KIDD BSL

British Heart Journal 33(Suppl): 1971; 81-87

Extracted Summary

The present study is an attempt to arrive at a clearer recognition of the true prevalence of ventricular septal defect and the various responses to it of the human body. There are a variety of figures given in the literature of the last 2 to 12 years on the incidence of spontaneous closure of the ventricular septal defect: (1) Keith, Rowe, and Vlad (1958), 10% closing; (2) Hoffman and Rudolph (1965), 24% closing; (3) Ash (1964), 15% closing; (4) Li et al. (1969), 22% closing.

The present report summarized 2 groups: (1) The 630 cases followed from infancy for an average of 7.5 years with an overall closure rate of 17%. The small defects identified clinically had the highest closure rate (26%). (2) Among the 295 cases followed through the teens into the twenties 4% closed spontaneously.

Perhaps equally important is to recognize early in life which cases are likely to close of their own accord, particularly in the first year or two of life.

Documentation of Spontaneous Functional Closure of a Ventricular Septal Defect During Adult Life

SCHOTT GD

British Heart Journal 35: 1973; 1214-1216

Extracted Summary

Spontaneous functional closure of a previously haemodynamically significant ventricular septal defect between the ages of 23 and 40 has been documented by serial cardiac catheterizations.

Selected Case Report

A congenital heart lesion was suspected in this man at the age of 4, when a cardiac murmur was found during routine examination. On two occasions, at the ages of 12 and 16, he was confined to bed for some months on account of an undiagnosed febrile illness, considered to be bacterial endocarditis.

At the age of 23, he was admitted for investigation, and examination was unremarkable apart from a systolic thrill and pansystolic murmur heard over the precordium, maximal at the left sternal edge. Radiographic screening of the chest showed prominent pulmonary arteries with increased pulsation, and hyperactive left and right ventricles. The clinical and radiographic impression of a ventricular septal defect was confirmed at cardiac catheterization: a 15% step-up in oxygen saturations at ventricular level, a pulmonary:systemic blood flow ratio of 2.5:1, and a left-to-right shunt of approximately 9 liters/minute were demonstrated. Intracardiac pressures were normal, and no abnormal communications could be crossed. Angiography was not performed.

He was seen again 17 years later, asymptomatic, at the age of 40. The signs on physical examination were unchanged, and the electrocardiogram and chest x-ray were normal. It was felt that the clinical findings were incompatible with the earlier investigations, and he was readmitted for a second catheterization. This showed no detectable step-up in oxygen saturations, and all intracardiac pressures were normal, apart from a right ventricular pressure of 38/0 mmHg recorded on only a single occasion, and an infundibular pulmonary gradient of 6 mmHg. Left ventricular angiography revealed a small aneurysm of the left ventricular wall at the site of the membranous septum, and a very small jet of contrast medium leaving the left ventricle from the centre of the aneurysm was seen. The cardiac output and pulmonary vascular resistance were normal.
Natural History of Ventricular Septal Defect

A Study Involving 790 Cases

Corone P; Doyon F; Gaudeau S; Guérin F; Vernant P; Ducam H; Rumeau-Rouquette C; Gaudeul P
Circulation 55(6): June 1977; 908-915

Extracted Summary

The development of 790 untreated patients affected by ventricular septal defect (VSD) has been the object of a 25-year study. Of these patients, 72% had had at least one catheterization; 13% had several. The mean observation interval is six years, and the average age at the latest data is 19.5 years. This study covers 4,717 patient-years.

Of the 499 cases with several clinical examinations, 71% remained stable. In 21%, changes suggesting some level of closure developed; in 7%, infundibular stenosis began to evolve and in 1% pulmonary vascular disease began to appear or became accentuated. These different rates are studied and discussed in relation to patients' age, VSD type, and various follow-up characteristics.

Spontaneous closure was observed; however, this study does not permit any evaluation of its absolute frequency, as one would have had to follow an entire population from birth to death. Nor could a true appraisal of the closure's frequency with age be made from our data; the time elapsed between two successive examinations varied widely and in some the interval was very long and prevented any reasonable estimate of the age when spontaneous closure took place.

The age characteristics of patients with spontaneous closure were looked at. The age at which the defect was last seen open was evaluated. This analysis will underestimate the age of closure, but may help in determining the chances a child of a particular age has of spontaneous closure. It appears that the younger the child, the greater his chances. Two of the VSDs did close relatively late, after the age of 16 and 17.

The size of the VSD plays a part in determining the chances of closure. While smaller VSDs are more likely to close, larger VSDs may also follow the same course: 58 out of the 215 VSDs in group I (27%) closed completely; 37 out of 133 VSDs in IIa (28%) closed completely (10 cases) or partially (27 cases); eight out of 82 VSDs in group IIb (10%) closed completely (one case proved by two catheterizations) or partially (seven cases). We have observed no closures among the VSDs in groups III and IV.

Ventricular Septal Defect: Late Spontaneous Closure

Rozza AMS; Schulze I; Keck EW

Extracted Summary

Twenty-five patients who had a proven isolated small ventricular septal defect (VSD) when they were children were re-examined 3 to 19 years later, at the age of 20-21 years. Spontaneous closure was diagnosed if the murmur had disappeared and all other cardiological findings were normal. This was so in nine patients. Six of them had previously been followed to eight years or more with the definite diagnosis of VSD. In the three others who had only been followed to the age of 3-5 years the time of closure, whether early or late, could not be assessed.
**Spontaneous Closure of a Coronary Artery Fistula in Childhood**

**MAHONEY LT; SCHIEKEN RM; LAUER RM**  
_Pediatric Cardiology 2(4): 1982; 311-312_

**Extracted Summary**

An infant with a symptomatic coronary artery fistula, documented by angiography, is presented. By age 5 years, clinical evidence of the fistula was no longer present. Repeat cardiac catheterization confirmed spontaneous closure of the coronary artery fistula. More information regarding the natural history of a coronary artery fistula is required before an elective surgical approach can be recommended in all asymptomatic children.

**Selected Case Report**

A 5-year-old female was first noted to have a heart murmur at age 5 months. When evaluated at age 8 months, a Grade III/VI continuous murmur was noted with maximal intensity at the lower left sternal border. The first heart sound was normal and the second heart sound was widely split. The liver was palpable at 3.5 centimeters below the right costal margin. Her chest x-ray demonstrated cardiomegaly and increased pulmonary vascular markings. The electrocardiogram showed a mean frontal axis of 60°, right ventricular hypertrophy, and possible left ventricular hypertrophy. At cardiac catheterization, oxygen saturations gave a calculated QP/QS of 1.4. The right and left heart pressures were normal. The systemic arterial pulse pressure was normal. An ascending aortogram demonstrated marked dilatation and tortuosity of the left circumflex and right coronary arteries. Angiographic contrast passed through this fistula to outline the body of the right ventricle.

Because of the mildly symptomatic nature of this defect, we elected to follow her medically with digoxin and endocarditis prophylaxis. After 2 years her heart size on chest x-ray was within normal limits. The digoxin was discontinued. The murmur decreased to an intensity of Grade II/VI and the electrocardiogram was normal. At age 5 years the murmur was no longer audible and the second heart sound split physiologically. An echocardiogram was normal and displayed normal septal motion.

A repeat cardiac catheterization was performed. There was no demonstrable step-up in oxygen saturation and no shunt was appreciated by dye curve. An aortic root angiogram showed the right coronary artery and left main coronary artery. The proximal portion of the left coronary artery showed evidence of residual mild dilatation and tortuosity and normal sized distal coronary arteries. No fistulous communication could be demonstrated. The left ventricle was normal in size and function. At her last clinical follow-up the electrocardiogram remained within normal limits.

**Spontaneous Complete Closure of a Congenital Coronary Artery Fistula**

**GRIFFITHS SP; ELLIS K; HORDOF AJ; MARTIN E; LEVINE OR; GERSONY WM**  
_Journal of the American College of Cardiology 2(6): Dec 1983; 1169-1173_

**Extracted Summary**

The first documentation is reported of spontaneous closure of a coronary artery to right ventricle fistula that was demonstrated initially in a 14-month-old boy. A heart murmur was first noted at 2 months of age. At examination, the boy was asymptomatic and his growth and development were normal. At 14 months of age, elective cardiac catheterization and angiography was performed. A small left to right shunt was detected. At 2 and 3 years of age the murmur was still present. Over the 4 year period after diagnosis, the characteristic continuous murmur gradually disappeared.

When the patient was 5 1/2 years of age, selective coronary arteriography showed normal coronary vessels and circulation. At subsequent examinations over the next two years, the boy was normal.
Six other cases of coronary fistula observed during the past 10 years are also reviewed. This study supports the rationale for clinical follow-up rather than obligatory surgical intervention in asymptomatic patients with a small shunt who have no evidence of myocardial dysfunction. (Permission to reproduce case report denied by author.)

**Spontaneous Closure of Coronary Artery Fistula**

**HACKETT D; HALLIDIE-SMITH KA**

*British Heart Journal 52(4): Oct 1984; 477-479*

**Extracted Summary**

A coronary artery fistula diagnosed in a 1-year-old girl closed spontaneously during childhood. This outcome has been documented in only three previous cases.

**SELECTED CASE REPORT**

A West Indian girl was referred in 1976 at the age of 1 year for investigation of an asymptomatic murmur, which had been discovered on a routine infant examination. Her medical history, birth and development, was unremarkable, and no murmurs had previously been noted. Examination at that time detected bounding arterial pulses and a systolic thrill with a grade 4/6 continuous murmur at the lower right sternal edge. The clinical diagnosis, based on the character and site of the murmur, was a coronary fistula. An electrocardiogram showed changes of biventricular hypertrophy, and a chest radiograph indicated minimal cardiomegaly and pulmonary plethora. Cardiac catheterization in January, 1977 (aged 1 1/2 years) confirmed a left to right shunt at the right ventricular level with a calculated pulmonary to systemic flow ratio of 1:4. Haemodynamic pressures in the right atrium, right ventricle, pulmonary artery, left ventricle, and aorta were all normal. Angiography of the aortic root showed a dilated proximal right coronary artery with a fistulous communication to the right ventricle just below the outflow tract. It was decided to manage this conservatively, and advice regarding prophylaxis against infective endocarditis was given.

During the period February, 1977 to February, 1979 the patient’s mother reported that the systolic thrill had disappeared. The patient remained asymptomatic. At the end of this period, when aged 4 years, she had no evidence of bounding pulses, systolic thrill, or any continuous murmur. Both the electrocardiogram and chest radiograph were normal. A treadmill exercise test (Bruce protocol) was negative for symptoms or electrocardiographic changes. Repeat cardiac catheterization in August, 1983 (at 8 years) again showed normal haemodynamic pressures, but no evidence of any left to right shunting could be detected. Angiography again showed a dilated right coronary artery ostium but no fistula was evident. A small right coronary artery distal to the previous fistula site was found.

**SUPPLEMENTAL REFERENCES**

**Coronary Artery Fistula**

The Spontaneous Disappearance of Interatrial Shunts in Infancy

**WATSON GH; DARK JF**

*Royal Society of Medicine. Proceedings 61(3): 1968; 300-02*

Spontaneous Near-Closure of Coronary Artery Fistula

**SHUBROOKS SJ; NAGGAR CZ**

*Circulation 57(1): Jan 1978; 197-199*
Anomalies of the Pulmonary Artery

Spontaneous Resolution of Pulmonary Stenosis

CHAN KC; CLARK D; GIBBS JL
International Journal of Cardiology 24: 1989; 375-377

Extracted Summary

Pulmonary stenosis diagnosed in infancy usually increases in severity with age or, if it is mild, may remain so for many years. We report the case of a child in whom moderately severe pulmonary stenosis in infancy appeared to completely resolve by the age of 8 years.

Selected Case Report

A 7-week-old girl, having presented with an asymptomatic murmur, was found to have the clinical signs of pulmonary stenosis. The chest radiograph was normal but the electrocardiogram showed evidence of right ventricular hypertrophy. Cardiac catheterisation at the age of 5 months confirmed the diagnosis, showing stenosis at both valvar and infundibular level. The right ventricular systolic pressure was 70 mmHg and an overall gradient of 50 mmHg was demonstrated across the right ventricular outflow tract. Angiography showed the pulmonary valve to be thickened and to dome during systole, suggesting partial fusion of the commissures. She was treated conservatively and remained well. By the time she was four, there had been a striking decrease in the intensity of the murmur and the electrocardiogram no longer appeared abnormal. Repeat cardiac catheterisation at this stage showed a right ventricular systolic pressure of 32 mm Hg, the outflow gradient having fallen to 14 mmHg. At outpatient review at the age of eight years, there were no audible murmurs. The heart appeared normal on cross-sectional echocardiography and Doppler ultrasound showed the peak flow velocity across the pulmonary valve, at 1.6 m/second, to be at the upper limit of normal.

Diseases of Blood and Blood Forming Organs

Anemias

Spontaneous Recovery of the Hemolytic Uremic Syndrome with Prolonged Renal and Neurological Manifestations

KAHN SI; TOLKAN SR; KOTHARI O; GARELLA S
Nephron 32: 1982; 188-191

Extracted Summary

A 3-year-old child had severe hemolytic-uremic syndrome with oligoanuric for 31 days and coma for 35 days associated with cortical blindness. She was treated with peritoneal dialysis and supportive care only, and recovered both neurologic and renal function. The spontaneous recovery of this patient, who had a particularly severe form of the hemolytic uremic syndrome, brings into question the necessity for the use of anticoagulation, antiplatelet agents, prostacyclin, and plasma exchange as treatments for the hemolytic-uremic syndrome.
A 3-year-old girl, D. L., developed diarrhea 4 days before admission. This was followed by severe and persistent vomiting. She then developed progressive lethargy and disorientation. A gradual decrease in urinary output was noted. On the evening of admission after a left focal motor seizure, she was seen in another hospital where she was given phenobarbital and fluids. The serum creatinine was 5.5 mg/dl (486.2 micro moles/l), hemoglobin 10.1 gm/dl and platelet count 41,000/mm³. She was then transferred to Rhode Island Hospital.

At admission she was comatose. Blood pressure was 130/86 mmHg, pulse was 150/minute and respirations were 20/minute. Temperature was 37°C., weight was 13.6 kilograms. Her skin was pale and the turgor was normal. Pupils were round, regular and responded to light; the ocular fundi revealed an exudative retinopathy. Her lungs were clear. Cardiac examination revealed a grade II/VI systolic murmur heard along the left sternal border. The liver was palpable 4 centimeters below the right costal margin. The patient responded to noxious stimuli, but there were no purposeful movements. Intermittent decerebrate posturing and hemiparesis were noted on the left. Babinski signs were present bilaterally.

At admission her blood glucose was 372 mg/dl (20.7 mmoles/l), BUN 156 mg/dl (55.7 mmoles/l), creatinine 6.2 mg/dl (548.1 micro moles/l), sodium 134 mEq/l (mmoles/l), potassium 5.1 mEq/l (mmoles/l), chloride 90 mEq/l (mmoles/l), and total carbon dioxide 20 mEq/l (mmoles/l). Calcium was 6.9 mg/dl (1.7 mmoles/l), phosphate 8.6 mg/dl (2.8 mmoles/l), uric acid 18.3 mg/dl (1.1 mmoles/l). LDH was 6,675 IU/l, SGOT 272 IU/l. Magnesium, ammonia, bilirubin, total protein, albumin, fibrinogen and lead levels were normal. Hemoglobin was 9.8 gm/dl with a microhematocrit of 27.3%. Reticulocyte count was 2.9% and platelet count was 31,000/mm³. White blood cell count was 9,000/mm³. Examination of the peripheral blood smear revealed numerous schistocytes, burr cells and helmet cells. Plasma fibrin split products titer was 1:20, prothrombin activity was 87% of normal control. Fluorescent antinuclear antibody was negative. Urine analysis revealed 3+ protein-urea, numerous red blood cells and no casts.

Spinal fluid revealed 45 red blood cells/mm³, 1 polymorphonuclear leukocyte/mm³ and 1 monocyte /mm³, a glucose concentration of 111 mg/dl (6.2 mmoles/l) and a total protein of 10 mg/dl. An EEG was consistent with metabolic encephalopathy and a right hemisphere focal seizure pattern. Echoencephalogram revealed no midline shift. A 99mTc glucoheptonate renal scan and an IVP with tomography failed to visualize the kidneys; a renal sonogram revealed 2 normal-sized kidneys. Biopsy of the kidney on the 18th day is described below.

The patient was initially treated with dexamethasone, phenytoin and intravenous fluids and peritoneal dialysis. Other than an initial brief rise to 150/100 mmHg, the blood pressure remained normal. Seizure activity continued intermittently through the first 8 hospital days. On the 7th hospital day large subhyloid hemorrhages were seen in both ocular fundi; these resolved by the 14th day. During the first 34 hospital days she remained comatose. On the 35th hospital day she began to demonstrate some purposeful movements; on the next day she was able to respond to verbal commands and began to talk. On the 40th hospital day it was first noted that she had cortical blindness. By discharge on the 47th hospital day the patient had shown marked improvement; however, she continued to have left-sided weakness, a broad-based gait, and ataxia. Her cortical blindness had improved to the point where she could recognize large shapes and objects.

Peritoneal dialysis was continued regularly until the 34th hospital day. This was complicated by a Staphylococcus aureus peritonitis on the 28th hospital day which was treated with gentamicin and oxacillin. She was anuric for 19 days and oliguric for 12 more days. At discharge her serum creatinine was 1.6 mg/dl (141.4 micro moles/l) and blood pressure was 120/80 mmHg. Five months after discharge the patient’s serum creatinine was 0.9 mg/dl (79.6 micro moles/l) and a 24 hour urine collection revealed 1.28 grams of albumin. Her microhematocrit had risen to 38%, platelet count was normal, blood pressure was 120/90 mmHg, and neurological examination was also normal. 1 year after discharge her creatinine was 0.8 mg/dl (70.7 micro moles/l), 24 hour urine protein excretion was 0.47 grams and creatinine clearance was 43 mm³/minute/1.73 m². According to her mother, her level of school performance, vision and family interaction were normal.

Renal Biopsy Findings: Light microscopic examination revealed patchy cortical necrosis. Interlobular arteries and arterioles showed marked fibrointimal proliferation and thrombosis and an occasional focus of fibrinoid necrosis without inflammatory reaction. There was deposition of fibrin within the walls of a few interlobular arteries and arterioles. In the less involved areas, the glomeruli showed accentuated lobulation, mild mesangial hypercellularity and thickened capillary walls. Occasional glomerular capillaries also showed fibrin thrombi. The interstitium showed edema and mild mononuclear infiltrate.

The immunofluorescent staining for fibrinogen revealed bright staining of arterial and arteriolar walls, capillary thrombi, and focal, segmental, bright deposits along the glomerular basement membrane and in the mesangium. Some glomeruli also revealed weak deposits of immunoglobulins (IgG, IgA, and IgM). On electron microscopic examination, the changes were consistent with those seen in microangiopathic hemolytic anemia; the most striking feature was the presence of large, pale, spongy subendothelial deposits in the glomerular capillaries.
Spontaneous Remission of Refractory Anemia with Excess of Blasts After Septicemia

DEHARO S; BIZET M; MONCONDUIT M; TILLY H; SARI F; PIGUET H
Annales de Medecine Interne 138(8): 1987; 663-664

Extracted Summary
The spontaneous evolution of an acute leukemia toward a complete remission is uncommon. Such remissions during the course of myelodysplastic syndromes are even more uncommon. We will report on a case of spontaneous regression of a refractory anemia with an excess of blasts (AREB) during a severe infectious episode. (Noetic Sciences translation)

Spontaneous Remission in Myelodysplastic Syndrome
A Case Report

BROWN ER; HEEREMA NA; TRICOT G
Cancer Genetics and Cytogenetics 46(1): May 1990; 125-128

Extracted Summary
A patient with refractory anemia with excess blasts (RAEB) and pancytopenia is reported. He was red blood cell (RBC) transfusion dependent. Karyotype analysis showed a complex cytogenetic abnormality consisting of loss of chromosome 7 and trisomy 8. Without cytotoxic treatment, his complete blood count (CBC) subsequently became normal. He is no longer transfusion dependent and repeat cytogenetic analyses are normal.

Coagulation Defects

Spontaneously Acquired Factor VIII Inhibitor in a Non-Haemophiliac Child

LABBE A; DUBRAY C; BEZOU MJ; TRAVADE P; COULET M
Acta Paediatrica Scandinavica 72: 1983; 621-623

Extracted Summary
A three-year-old girl who had for two months suffered bruising after minimal injury was admitted because of diffuse ecchymoses and a large haematoma hindering elbow movement. These symptoms were attributable to the development of antifactor VIII inhibitor. No definite etiology was evident despite repeated immunological investigation. Although the inhibitor still persisted at high levels after two years, no further haemorrhage occurred, excepted haematomas three months after the onset of symptoms, in association with mumps.

Clinical History: A three-year-old girl was admitted in November, 1980 because of bruising. She had been in good health until two months prior to admission when ecchymoses appeared frequently after minimal injuries. Both the patient's and the family history were negative for autoimmune disorders or haemorrhagic diathesis. There had been no exposure to toxins or drugs except penicillin, given four months before. On admission, clinical examination showed extensive ecchymoses on the limbs, small bruises over the trunk and a large subcutaneous haematoma on one elbow. There was no lymphadenopathy or hepatosplenomegaly, nor fever. The large subcutaneous haematoma resolved after a week. The child has been followed for two years. No other bruising or bleeding has occurred.
Laboratory Data: Haematological findings were as follows: haemoglobin 140 gm/l, white blood count 12.4 x 10^9/l, differential count normal, no eosinophilia, platelet count 280 x 10^9/l. Serum immunoglobulins (IgG, IgM, IgA, IgE) and levels of C3 and C4 were normal. E rosettes were 70% and EAC rosettes were 18% which are normal values for age. No antinuclear, smooth muscle or mitochondrial antibodies were detected. Viral serological investigations for adenovirus, respiratory syncytial virus and Epstein-Barr virus were negative. The IgE antibody activity against penicillin G and V was determined by the radioallergosorbent test (RAST) using ^125^I, labelled anti DE2. The results expressed in PRU/ml (Phadebas RAST units, Pharmacia Diagnostics) 0.3 PRU/ml was considered positive.

Circulating Anticoagulant Due to Factor VIII Deficiency With Spontaneous Remission

*Case Report*

GETZEN JH
*Southern Medical Journal 60(4): Apr 1967; 378-381*

*Extracted Summary*

This is a presentation of a 41-year-old menstruating woman who developed evidence of circulating anticoagulants and was later found to have inhibitory qualities against factor VIII. She maintained this defect for a period of 5 years and had a spontaneous remission. During the course she had several life-threatening hemorrhages, either from blood loss or involvement in a vital area such as the chest, paratracheal region and intralumenal obstruction. Initially, a number of agents were given as a therapeutic trial, including steroids, all of which did not produce any significant alteration in her coagulation defect. With the judicious use of blood transfusions, mainly to replace lowered blood volume, she was able to maintain her same blood group and never developed a serious blood reaction.

Her last serious illness was characterized by congestive heart failure, hypotension and at one point she was considered in extremis, but recovered and was discharged without evidence of recurrent bleeding. Her subsequent coagulation studies have now remained entirely normal as of this date.

Lupus Anticoagulant and Acquired Prothrombin Deficiency During Viral Disease in a Child

*Spontaneous Recovery*

HOUBOUYAN L; ARMENGAUD D; LEROY B; ROUSSI J; GALLE JP; GOGUEL A
*Archives Francaises de Pediatrie 41(6): Jun-Jul 1984; 417-420*

*Extracted Summary*

A 2-year-old female developed an acute bleeding diathesis related to a profound, isolated and acquired prothrombin deficiency; evidence for a lupus anticoagulant was also demonstrated. This association of hypoprothrombinemia and lupus anticoagulant, rarely reported, was previously considered to be rather specific for SLE. This case report demonstrates that these coagulation disorders may present as an acute form, in viral diseases of the child, with spontaneous and quick recovery. Specific characteristics of the biological coagulation defects, namely those related to the low factor II, are discussed.
A Neonate with Down’s Syndrome and Transient Abnormal Myelopoiesis: Serial Blood and Bone Marrow Studies

**Nagao T; Lampkin BC; Hug G**
*Blood 36(4): October 1970; 443-447*

**Extracted Summary**
Observations were made of the blood and bone marrow of a male infant with Down's syndrome during the first year of life. At 4 days of age there were 36,500 myeloblasts/mm³ in the blood and 10.8% myeloblasts in the marrow. Initially it appeared the patient had acute myeloblastic leukemia. However, the clinical course and kinetic and electron microscopic studies of his bone marrow cell population indicate he did not have acute leukemia.

Angio-immunoblastic Lymphadenopathy with Spontaneous Remission in an Over 80-Year-Old Female Patient

**Zilotto G; Miotto P**
*Giornale di Gerontologia 28(8): 1980; (Recd 1981) 541-552*

**Extracted Summary**
A case of angio-immunoblastic lymphadenopathy (AIL) in an over 80-year-old female patient is reported. The onset of the disease was characterized by general malaise, pruritus and macupapular rash on the legs, focal lymphadenopathy, strong polyclonal gammopathy, obliteration of the lymph node architecture due to a proliferation of lymphocytes, plasma cells, immunoblasts, small vessels, and deposit of amorphous PAS-negative material. The course was favorable with spontaneous remission of the adenopathy, progressive normalization of the protein disorder, lymph node cellular depletion with the persistence only of the lymphocytes. This disease, which may be caused by a viral infection, is considered a benign type of AIL where the immunologic disorder is only temporary.

Angio-immunoblastic Lymphadenopathy with Dysproteinemia

**Fiorillo A; Pettinato G; Raja V; Migliorati R; Angrisani P; Buffolano W**
*Cancer 48(7): Oct 1 1981; 1611-1614*

**Extracted Summary**
This is the first known report of a case of angio-immunoblastic lymphadenopathy, with dysproteinemia (AILD) in childhood which evolved toward a spontaneous remission. The disease had an acute onset with generalized lymphadenopathy, hepatosplenomegaly, high-grade fever and polyclonal hypergammaglobulinemia. The lymph nodes met all of the histologic criteria required for diagnosis as established by Frizzera et al. It is emphasized that AILD should be taken into consideration in the differential diagnosis of lymphadenopathy in childhood.
Spontaneous Remission of Agnogenic Myeloid Metaplasia And Termination in Acute Myeloid Leukemia

RAGNI MV; SHREINER DP

Archives of Internal Medicine 141(11): Oct 1981; 1481-1484

Extracted Summary

Acute myeloid leukemia developed in two men with well-documented agnogenic myeloid metaplasia 49 and 27 months, respectively, after initial diagnosis. Both men had spontaneous hematologic remissions of myeloid metaplasia 22 and 16 months prior to leukemic transformation. Neither patient received any precedent radiation or cytotoxic chemotherapy. The remissions and subsequent transformation to acute leukemia were documented by bone marrow aspiration, biopsy, and reticuloendothelial marrow scans, as well as in-vitro cell cultures and cytogenetic study.

A review of the literature revealed nine other patients who had conversion to acute leukemia in the absence of preceding radiation or cytotoxic chemotherapy. However, the present report seems to be the first describing spontaneous hematologic remission prior to transformation to leukemia.

A Case of Angio-immunoblastic Lymphadenopathy Associated with a Long Spontaneous Remission, Retrobulbar Neuritis, A Clonal Rearrangement of the T-Cell Receptor Gamma Chain Gene and an Unusual Marrow Infiltration

MATTHEWS JH; SMITH NA; FORONI L

European Journal of Haematology 41(3): Sep 1988; 295-301

Extracted Summary

A man with angio-immunoblastic lymphadenopathy entered a spontaneous remission that lasted 18 years, ending in retrobulbar neuritis followed by a generalized relapse. After another remission was induced by prednisolone he developed marrow infiltration with a lymphoid cell of novel surface-marker phenotype. His lymph node cells demonstrated a clonal rearrangement of the T-cell receptor gamma chain (TcR gamma) genes whereas the TcR beta genes were heterogeneously rearranged. The marrow cells did not contain the rearranged TcR gamma gene. He died of pulmonary emboli 21 years after his original presentation.

We propose a model whereby the original event was the development of a lymphoid stem cell clone with no TcR gene rearrangement. Subsequently, a sub-clone bearing the TcR gamma gene rearrangement proliferated in the lymph nodes and further heterogeneous rearrangement of the TcR beta genes occurred within this subclone. Proliferation in the marrow of the original clone finally supervened.

Supplemental references

Diseases of Blood and Blood Forming organs

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Part Two: Diseases Other Than Cancer