A C A S E
OF
TRANSPPOSITION OF THE GREAT VESSELS
OF THE HEART.

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TRANSPPOSITION of the great vessels of the heart is sufficiently rare to warrant me in submitting to the Fellows of this Society the particulars of the following case :

W. L—, aged two years and four months, came under my care August 18th, 1858. The following history was given by his mother :—She has had four children. Of these, one died from hooping-cough, another from inflammation of the bowels, the third survives, and is strong and healthy. The subject of the case was born at the ordinary period, a remarkably fine, healthy boy. Two or three days subsequently he became jaundiced, but within a fortnight had regained his colour, and remained perfectly well, a strong, plump child, differing in no respect from other children until the completion of his third month. After this time it was observed that the lips, fingers, and toes turned blue. The breathing also became difficult, especially under the influence of anger. The child was considered remarkably passionate. Cough additionally supervened. Teething

began at the ordinary period, and was completed without difficulty. This peculiarity, however, was remarked—that in both jaws the back teeth were earliest cut. The child appeared to be unusually sensitive to cold. The mother particularly mentioned the unusual intelligence of the child. There was, however, so much weakness of the lower limbs that it could never support its feet upon the ground; they invariably gave way. With the exception of occasional diarrhœa, all the remaining functions seemed naturally performed. Latterly it has begun to lose flesh, and has, at times, a difficulty in sucking.

The child appeared, at the first glance, to be sinking from an advanced stage of pulmonary disease. The face was cold and pallid; the eyes prominent; the conjunctivæ dark. The lips, tongue, and interior of the mouth were of dark purple colour. The fingers and toes cool and clammy, were clubbed, and of leaden hue as far as the second phalanges. The impulse of the heart was extensively visible. Marked pulsation also existed at the epi-sternal fossa. No *fremitus* could be detected over the cardiac region. The sounds of the heart were apparently perfectly normal, excepting that the second sound was unusually loud and marked towards the third left costo-sternal cartilage. Considerable dyspnœa existed, but the lungs offered no definite physical signs. Those already mentioned scarcely ever altered in any important particular beyond paroxysmal increase, although the case was very constantly under observation.

The child, during the last four months of its life, became gradually more emaciated. It suffered also from increasing difficulty of breathing. The mother thought at this period that it was palsied in the lower limbs. Eventually the belly and feet began to swell, and during the latter weeks the body generally was covered with petechiæ. Death occurred, apparently, from exhaustion in the November following, the child having attained the age of two years and eight months.

The body was examined a few hours after death—the

head was not allowed to be opened. The discoloration of the lips, fingers, and toes still remained, although diminished in intensity. Upon removing the sternum, the thymus gland was seen still very large. The heart, distended with blood, occupied a considerable portion of the chest. The pericardium was extensively and very intimately adherent to the left pleura, the visceral and parietal layers of which were so closely united that the heart was firmly held in position. The left lung, concealed by the heart below the second and third ribs, was unexpanded in its lower two thirds, and had probably remained so from birth. The right lung, though somewhat pushed aside by the heart, was healthy. The liver was very large, and of dark purple colour: it occupied both the hypochondria. The remaining abdominal viscera were congested, but in other respects apparently unchanged. Moderate effusion existed in the peritoneal sac.

Upon examining the preparation of the heart and vessels, purpuric maculæ, of varying size, are visible upon the anterior aspects of the aorta and pulmonary artery. The same appearance exists also upon the posterior surface of the right ventricle. The heart is somewhat altered in shape, being broader at the base than usual. Its weight, as nearly as can be ascertained, allowing for the attachment of the vessels, is between three and three quarters and four ounces. It measures nearly 3 inches in the transverse, and nearly 2·2 inches in the vertical direction. The apex is very obtuse, or rather of semicircular outline, formed nearly equally by right and left ventricles; the left slightly predominating. The aorta arises from the fleshy part of the base of the right ventricle, at its left portion, where it presents itself in front, being quite uncovered by the pulmonary artery, and is situate between this vessel and the right auricle. It measures above the valves ·6 inch, transverse diameter. At the descending portion of the arch, anterior to the origin of the arterial duct, the vessel is slightly contracted, measuring at this spot, as well as before the innominate trunk, ·43 inch. The aorta gives off its coronary branches, very large, in the ordinary manner. The arteries

of the arch also arise without any irregularity either in their number or position. From the upper and posterior aspect of the vessel, just beyond the arch, five small arteries, of the usual calibre, arise, probably bronchial, and œsophageal or thymic. Posteriorly the intercostal branches, also of normal calibre, are given off in the ordinary manner. The semilunar valves of the aorta are three in number, and manifestly competent, being normal in size and structure. The pulmonary artery is very large, measuring just above the valves one inch in diameter, and immediately at the point of division 1·1 inch. It springs from the anterior part of the left ventricle, the aorta being close to it on its right, and the tip of the left auricle on its left side. It divides, in the usual manner, into its two branches. The branch to the right lung measures transversely ·6 inch; that to the left lung ·55 inch. The three semilunar valves of the artery are of correspondingly large size and strength. The ductus arteriosus is completely closed. The fibrous cord representing it extends from the upper side of the pulmonary artery, close to the origin of its left branch, to the under side of the aorta, a little beyond the origin of the left subclavian artery. The right auricle is dilated and hypertrophied. Its walls measure at the thickest part, close to the entrance of the superior cava, ·12 inch. The size of its cavity, from the tip of the auricle to the entrance of the inferior cava, is about 2 inches; from the auriculo-ventricular junction to the edge of cavity between the veins, 1·2 inch. The foramen ovale is largely patent when stretched, admitting a cone, ·45 inch; or, with ease, the tip of the little finger. There are distinct traces of the remains of the valve of the foramen. The superior and inferior cavæ enter the right auricle in the usual way. The position and size of the coronary and azygos veins are normal. The right ventricle is greatly hypertrophied, and its capacity diminished. The thickness of the walls at the base varies; the right wall measures ·5 inch; left wall close to the septum and at attachment both of aortic and mitral valves, ·7 inch; anteriorly, at the origin of aorta, ·25 inch. A very strong, fleshy column, ·25 inch by ·45 inch, from which the prin-

cipal papillary muscle arises, forms a kind of imperfect septum in the ventricle; midway, between apex and base, the wall is of uniform thickness, measuring $\cdot 33$ inch. The left auricle is dilated; its diameter, from the base of the ventricle to the superior boundary, is $1\cdot 20$ inch; and about the same diameter from the inter-auricular septum to the tip of the auricle. The pulmonary veins, four in number, are normal in situation, and equal in size, emptying themselves as usual into the left auricle. The left ventricle is in a state of dilated hypertrophy. At the base, close to the septum, the right wall measures $\cdot 37$ inch; the left wall, $\cdot 23$ inch; anteriorly, close to the pulmonary artery, 1 inch $\cdot 05$; midway, between apex and base, $\cdot 28$ inch. The principal papillary muscle in the left ventricle is larger than that of the right; the latter is round, and measures $\cdot 15$ inch; the former irregularly square, $\cdot 30$ inch diameter. The ventricular septum is perfect; its diameter is about $\cdot 40$ inch. Both tricuspid and mitral orifices are manifestly competent. The right auriculo-ventricular orifice is oblate in form, and measures $\cdot 75$ by $\cdot 4$ inch. The tricuspid valve is of the usual form. The left auriculo-ventricular orifice is also oblate, measuring $\cdot 8$ by $\cdot 4$ inch. The mitral valve differs materially from the usual form, being really tricuspid. Its anterior cusp being attached to both of the posterior pulmonic valves, and to the wall at the base of the cavity, close to the auricular portion; the inner cusp is attached mainly to the septum; and the outer and posterior cusp mainly to the base of the ventricle. Notwithstanding this peculiarity of the mitral valve, the perfectly normal condition of the tricuspid valve in the right ventricle shows that the ventricles themselves were not transposed.

It will be seen from the above description that the only possible communication between the two sides of the heart, and consequently between the systemic and pulmonic circulation, was through the foramen ovale.

Remarks.—There are certain features in this case more or less common to the entire collection of cases of transposition of the vessels, which appear worthy of notice. Such are the

enlarged state of the thymus gland ; the imperfect expansion of the left lung ; pleuro-pericarditis ; the tendency to exudation of black blood, and the condition of the nervous centres, *i. e.*, the apparent integrity of the cerebral functions, coexisting with diminished function of the spinal axis ; and, eventually, the failing power of the sympathetic system as evidenced in the impaired nutrition of the body. And these points are all, more or less, inseparably connected with malformation of the heart generally.

An enlarged thymus gland has been noted by Duretus, Sandifort, Meckel, Gintrac, Cailliot, and Burns, and supposed by them to exercise a compensating influence.

So many cases of unexpanded and of imperfectly developed left lung have occurred, not only in transposition of the vessels, but in other malformations of the heart, that there would seem some casual relation between the two conditions. For original observations on this point, see Meckel, 'Handbuch der Patholog. Anat.,' B. i, s. 176 ; and Heyfelder, 'Studien im Gebiete der Heilwissenschaft,' B. i, s. 233.

The comparative frequency of pleuro-pericarditis would also justify the inference that malformed heart confers a special predisposition to the occurrence of such complication.

Exudation of black blood, either in the form of slight hæmorrhage from the mucous membranes, or in that of petechiæ and purpuric maculæ, or of aphthæ, is a very constant phenomenon. Heyfelder (*op. cit.*, B. i, s. 228) has bestowed particular consideration upon this subject.

With respect to the cerebral functions, the admixture of black blood, even to a great extent in the nutrient vessels of the brain, does not appear in many cases to modify, in any perceptible degree, their normal manifestations. Indeed, some physicians, Lentin ('Beiträge,' B. ii, s. 70), have particularly noted the excellence of the intellect in the cyanotic patient. Sachse also records the instance of a cyanotic youth, at the age of twenty years, pursuing his studies at the university. Indeed, it conforms to our ordinary experience that cyanotic adults are but little, if at all, below the average standard of intelligence.

But while the cerebral functions remain in general un-

modified in transposition of the vessels, in nearly all the minutely recorded cases there has existed a diminished power of voluntary motion ; in some cases shown by a simple inability to execute properly the healthy motions of the lower extremities ; in others by the existence of actual paralysis of either upper or lower limbs. In every case, the patients have had an almost instinctive dread of voluntary movement, in consequence of such movement determining convulsive or syncopal paroxysms.

The sympathetic system appears, in the cases that have survived for any period, to have remained for a time without serious compromise. The growth of the body has proceeded normally, and the various functions have been naturally performed. But after a time, emaciation and general perverted nutrition have supervened, terminating in gradual exhaustion of the vital powers.

The collated cases of transposition of the great vessels of the heart amount, as nearly as the author can ascertain, to about forty-two in number. It is possible, however, that one or two of the cases may have been twice quoted in consequence of appearing at one time in the name of the observer, at another in that of the collator. To facilitate reference, they are arranged in a triple series ; commencing with the most complex cases, and ending with those in which the transposed vessels were associated with the smallest deviation from the normal condition of the heart.

The first series includes those cases in which transposition of the vessels is associated with arrested development and malposition of the trunks of the main vessels ; structural changes in the various orifices of the heart ; deficiency of the ventricular septum ; transposition of the ventricles and their valves.

The second series comprises the cases in which transposition of the vessels is accompanied by patency, both of the foramen ovale and of the ductus arteriosus.

The third series contains the few examples in which transposition of the vessels occurs with simple patency alone of the foramen ovale.

SERIES I.

	Observer.	For. ov.	Sep. vent.	Duct. arteriosa.	Duration of life.	Remarks.	Reference.
1	Stedman	Open	Open	Closed	7 m. 8 d.	Cyanosis after fourteen days. Right lung largest, left compressed. Pericarditis. Irreg. dist. of vessels of aortic arch. Pul. art. dilated above its mouth. Edema and ascites	Lancet, 1841-2.
2	Bock	—	Open	Open	8 m.	Aorta arose from R. v. Pul. art. over sep. vent.	Freidberg, op. cit.
3	Meckel	Open	Open	Open	2 d.	In a lamb	Tab. Anat. Path.
4	Wilkinson King	Open	Open	Closed	2 y. 9 m.	Cyanosis. Pul. art. wide; aorta small below duct; only two veins entered left aur. Never walked	Monthly Journ. of Med. Sc., vol. iv.
5	Nasse	Open	Open	—	19 y.	Cyanosis. Convulsions	Leichen offnungen 1ter Theil, 1821.
6	Meckel	—	Open	—	—	Bicephalous calf	Tab. Anat. Path.
7	Thore	Open	Rud.	—	11 d.	No cyanosis	Arch. Gen., s. iv, t. 1, 1843.
8	Otto	Open	Absent	Absent	7 m. foetus	Pul. art. too small	Selt. Beobachtungen.

9	Mauran	Sep. abs.	ditto	Br. of P. a.	—	Pul. art. impervious at its origin	Freidberg, op. cit.
10	Breschet	ditto	ditto	Abnormal	—	Duct. art. was a long, slender canal between right branch of P. a. and right subclav. P. a. contracted	Otto. Selt. Beobach.
11	Lawrence	ditto	ditto	—	—	P. a. contracted	—
12	Langstaff	Closed	—	Nor. width	10 w.	P. a. branch of descending aorta	Lond. Med. Rev., v. 4.
13	Kreysig	ditto	—	—	—	P. a. branch of descending aorta. Tricuspid orifice closed	Krank. des Herz, B. iii.
14	Valleix	Small chink	Open	—	8 d.	Inver. of viscera. Tricuspid orifice closed	Arch. Gen., s. ii, t. 8.
15	Müller	Three small chinks	Open	—	1 y.	P. a. contracted	Horn's Archiv, 1822.
16	Farre	Open	Absent	Br. of P. a.	79 h.	P. a. imperfect. Aorta arises over sep. vent.	Malf. of Heart.
17	Raoul-Chassinat	—	Open	Open. Crow quill into left P. a.	12 d.	P. a. rudimentary. Defective right aur. and vent.	Friedberg, op. cit.
18	Martin	Wide open	Open, two lines	Nearly obliterated	10 w.	Common aur. and vent. from which the great vessels transposed arose. No cyanosis	Müller's Archiv, s. 222, 1859.
19	Farre	Imperfectly closed; val. perforated	—	Contracted	5 m.	P. a. contracted. Cyanosis	Malf. of Heart.

	Observer.	For. ov.	Sep. vent.	Duct. arterios.	Duration of life.	Remarks.	Reference.
20	Boyer	Open	—	Closed in part	2 m.	P. a. connected with right vent. Aorta arose above aperture in septum, and communicated with both vent. Cavæ entered left and pul. veins right aur. viscera, generally reversed	Peacock, op. cit.
21	Worthington	—	—	—	—	Similar case	Idem.
22	Hess	Closed	—	Probably open	8 m.	Numerous abnormal conditions, occasional lividity only. Male	Idem.
23	Burkart	—	—	—	—	—	De Monst. Notab. Friburg. 1825.
24	Johnson	—	—	—	2 m.	Male	Amer. Journal of Med. Sc., new ser., vol. xx.
25 and 26 27	Stolz Hesse	— —	— —	— —	— —	{ Two cases. 1st case, infant lived five days; 2nd case, thirty-seven days Probably, defective aur. and vent.	Gaz. Méd. de Strasbourg, quoted by Peacock, op. cit. Freidberg, op. cit.
SERIES II.							
28	Baillie	Slightly closed	—	Size, large enough to admit crow-quill	2 m.	Cyanosis. No dyspnoea. Heart of natural size. Child healthy 1st.	of Morbid Anatomy, Fasc. 1st.

29	Ducrest	Open	Perfect	Open	10 h.	Cyanosis.	Well formed.	Female	Archiv Gén. 3me et nouvelle série, t. 9, 1840.
30	Guy's Museum, 1392 ⁴⁰	Open	ditto	Open	—	—	—	—	—
31	Walshe	Open	ditto	Open	10 m.	Cyanosis.	Ventricle transposed.	Male	Med.-Chir. Trans., 1842.
32	Tiedemann	Wide open	ditto	Open, largely	12 d.	—	—	—	Zeitschrift, 1825.
33	Kell	Slightly open	ditto	Pretty wide	35 h.	—	—	—	Diss. Würzburg, 1825.
34	Ogier Ward	Open	ditto	Open	18 d.	Cyanosis from birth.	Dyspnoea. Difficulty in sucking. Lungs engorged and imperfectly expanded	—	Peacock, op. cit.
35	Coligny	Nearly closed	—	—	—	—	—	—	Pize, op. cit.
36	Meyer	Open, two lines	ditto	Open	1 m.	—	—	—	Virchow's Archiv, B. xii, 4, 5 Heft.
37	Beck	Open	ditto	Open	75 h.	—	—	—	Archiv für Phy. Heilkund, B. v, 1856.
38	Ribes	Open	—	—	3 y.	Left chamber contracted	—	—	Pize, op. cit.
39	Duges	—	—	Open	—	—	—	—	Jour. Gén. de Méd., vol. ci, p. 88.

Some of the cases quoted in the first and second series may be imperfectly classified, in consequence of the extremely meager details furnished by the writers quoting them; frequently a bare mention of the case as one of transposition being alone found. These cases the author has not been able to consult in the original records.

SERIES III.

(For further details, see Table at pp. 206, 207.)

	Observer.	For. ov.	Sep. vent.	Duct. arterios.	Duration of life.	Remarks.	Reference.
40	Gamgee ¹	Open largely	No mention	No trace	15 w.	Inver. of viscera. Cyanosis after some days; irreg. dist. of vess. of arch; pulmonary artery relatively large; muscular debility; aphthæ	New Eng. Jour. of Med. and Sur., vol. iv.
41	D'Alton	ditto	Perfect	Closed	1½ y.	Cyanosis beginning of third month. Pulmonary artery very large	Dissert. de Cyanopath., 1824.
42	Wistar	ditto	ditto	ditto	2½ y.	Pulmonary artery large. Cyanosis	Sys. of Anatomy, vol. ii, p. 75.
43	Author	ditto	ditto	ditto	2 y. 8 m.	Ditto. Cyanosis	—

¹ Although the case of *Gamgee* was conjoined with transposition of the viscera and cardiac ventricles, it is included in this series, as it conforms to the cases therein comprised in most particulars. The condition of the ventricular septum not being named, it must be inferred that it presented no abnormal condition.

The cases comprising this last series, though smallest in number, yet offer the fewest disturbing elements. In an anatomico-physiological sense, their characteristic consists, as stated in an open foramen ovale, associated with transposition of the vessels. The arterial duct is closed; no imperfection of the ventricular septum, or change, so far as can be learned from their detail, either in respect of size or origin of other vessels, compensating for such closure.¹ They are four; those of Gamage, Wistar, D'Alton, and the author. The following table shows how perfectly they coincide in every important particular; at least in all such as have been noted by the observer.

¹ It is hardly necessary to observe how carefully the branches given off by the thoracic aorta below the arch should be examined, not only in cases of transposition of the vessels, but in those of malformation of the heart generally, with the view to determine how far they could have exercised a compensating influence. Chevers ('Morbidity Conditions of the Pulmonary Artery,' p. 18) has already insisted upon this point. In some cases, the vicarious action of these vessels is shown beyond doubt. In an instance of obstructed pulmonic orifice recorded by Gerstel (Friedberg, *op. cit.*, p. 132), the bronchial arteries were so greatly enlarged, and as it were varicose, as to form an extensive network.

The manner in which the vessels arise from the arch should also be noted, inasmuch as several cases of transposition were accompanied with irregular origin of the vessels (Walshe, *op. cit.*). In D'Alton's case the origin and distribution of these vessels were normal; the innominate trunk, however, he mentions as being short. In the author's case these vessels were normal.

TABLE.—*Detailed account of the Cases in Group III, in which there was Transposition of the Great Vessels, the Foramen Ovale alone being open.*

GAMAGE's case.	WISTAR's case.	D'ALTON's case.	The AUTHOR's case.
<i>Female</i> .—Lived fifteen weeks.	<i>Male</i> .—Lived two years and six months. Average stature.	<i>Female</i> .—Lived one year and six months. Average stature.	<i>Male</i> .—Lived two years and eight months. Average stature.
Healthy until second month, but weak in limbs.	—	Healthy until third month.	Healthy until third month.
Cyanosis from birth, but much increased after second month.	Cyanosis, advent not stated.	Cyanosis beginning of third month.	Cyanosis end of third month.
—	—	Dentition easy.	Dentition easy.
Distressing cough and dyspnoea.	Dyspnoea.	Cough and dyspnoea.	Cough and dyspnoea.
Cyanotic tint deepening under effort.	Cyanotic tint deepening under slight dyspnoea, emotion, or effort.	Cyanotic tint deepening under emotion.	Cyanotic tint deepening under emotion.
—	—	Sensitive to cold.	Sensitive to cold.
—	—	Increased cardiac impulse.	Increased cardiac impulse.
—	—	Occasional diarrhoea.	Occasional diarrhoea.
Cerebral functions impaired (?).	—	Cerebral functions normal.	Cerebral functions normal.
Voluntary motion greatly impaired; right arm paralysed; eyes glassy.	Voluntary motion impaired.	Voluntary motion impaired.	Voluntary motion impaired.

_____	_____	_____	Latterly, difficulty in sucking.
_____	_____	_____	Emaciation.
Aphthæ over mouth and nates.	_____	_____	Petechiæ over body.
Severe convulsive attacks.	_____	_____	No convulsions.
_____	_____	_____	Edema and ascites.
Foramen ovale open largely.	_____	_____	Death from exhaustion.
_____	_____	_____	Foramen ovale open largely.
_____	_____	_____	Valve existing.
Ductus arteriosus seemed never to have existed.	_____	_____	Ventricular septum perfect.
Pulmonary artery large, relatively.	_____	_____	Ductus arteriosus closed.
Heart generally distended.	_____	_____	Pulmonary artery large.
Entrance of veins normal.	_____	_____	Heart dilated and hypertrophied.
_____	_____	_____	Mitral valve tricuspid.
_____	_____	_____	Branches of aortic arch normal.
_____	_____	_____	Entrance of veins normal.
_____	_____	_____	Left lung unexpanded in part.

For the general principles of arrangement and tabulation, and for much most valuable knowledge respecting the proximate cause of the malformation and irregularities of the large vessels generally, the author is indebted to the following authorities :

- Sandifort, 'Obs. Anat. Patholog.,' vol. iii, liber 1—3.
 Meckel, 'Handbuch der Patholog. Anat.,' Band i.
 Otto, 'Pathol. Anat.,' p. 297.
 Farre, 'Essay on the Malformation of the Human Heart.'
 Ecker, 'Beschreibung einig. fäll. von anorm. Commun.,' 1839.
 Albers, 'Atlas der Patholog. Anat.'
 Burdach, 'Traité de Physiologie,' t. iii, p. 502.
 Geoffrey St. Hilaire, 'Histoire des Anomalies,' &c., t. iii, p. 562.
 Cruveilhier, 'Anat. Patholog.,' t. ii, p. 474.
 Deguise, 'De la Cyanose Cardiaque,' &c., Thèse, 1843.
 Pize, 'Consid. sur les Anom. Card. et Vascul.,' Thèse, 1854.
 Gintrac, 'Sur la Cyanose,' 1824.
 Kürschner, 'Comment. de Cord. &c.,' 1837.
 Stille, "On Cyanosis," 'Amer. Jour. of Med. Scien.,' 1844.
 Freidberg, 'Die angeb. Krank. des Herz.,' &c., 1844.
 Kreysig, 'Krank. des Herz.,' B. iii.
 Chevers, 'Morbid Conditions of the Pulmonary Artery,' 1851.
 D'Alton, 'Dissert. de Cyanopath.,' &c., 1824.
 Nasse, 'Archiv für die Physiol. Reil und Autenrieth,' B. x.
 Walshe, 'Med.-Chir. Transactions,' vol. xxv.
 Peacock, 'Malformations of the Human Heart,' 1858.
 Meyer, 'Virchow's Archiv,' Band xii, 4tes und 5tes Heft, 1857.
 Turner, 'Brit. and For. Med. Quart.,' July and October, 1862.
 Wood, 'Path. Trans.,' vol. x, 1859.

Allen Thompson, "Case of right Aortic Arch.," 'Glasgow Med. Journ.,' April, 1863.

And the Handbooks of Rokitansky, Craigie, Engel, and Förster.

DESCRIPTION OF PLATES IX, X.

The plates represent a heart in which there is transposition of the great vessels, the foramen ovale being open.

- A. External view.
- B. Internal view.
- C. Posterior view.
- E. Right ventricle, from which the aorta takes its origin.
- L. Left ventricle, from which the pulmonary artery takes its origin.
- M. Mitral valve, which presents three cusps.
- T. Tricuspid valve.
- O. Foramen ovale.

B

Harvey O. Smith del. G.H.F.

C

W West. imp.